Experimental Dermatology

Abstracts of the 8th European Hidradenitis Suppurativa Foundation (EHSF) Conference, 6-8 February 2019, Wrocław, Poland

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ABSTRACT

PLENARY LECTURES

001 PL01 - oral session | Biomarkers of hidradenitis suppurativa/acne inversa

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The detection of biomarkers as reliable outcome parameters of the treatment of inflammatory diseases represents a major need, especially in clinical studies. A set of core outcome domains has therefore been identified. The current instruments to describe these domains may be clinical, such as the Sartorius or the IHS4 score or patient reported outcome measures or mixtures thereof. Ideally, they may also be biochemical or biophysical measures. In HS, specific biochemical or biophysical biomarkers have barely been detected until now, with c-reactive protein being the only one routinely reported in spite of its limited range in active HS. Other biochemical markers, such as interleukin (IL)-6, IL-17, sIL2-R, lipocalin-2, mi-RNA's, complement, chitinase-3-like protein and S100A8/A9 have been explored in smaller studies. A current own work has detected potential key regulators of HS pathology (DEFB4, MMP1, GJB2, PI3, KRT16, MMP9, SERPINB4, SERPINB3, SPRR3, S100A12, S100A7A, KRT6A, TMPRSS11D), which regulate innate immunity pathways in epithelial and lipids producing cells, involving glucocorticoid receptor signalling, atherosclerosis signalling, inhibition of matrix metalloproteases, HIF1α signalling and IL17A signalling. On the other hand, non-invasive and radiation-free clinical biomarkers, such as high resolution ultrasonography, long-wave medical infrared thermography, standardized clinical photography and their combinations can identify the true extent of lesions in HS, which may be of use both in the preoperative planning and in clinical follow-up of medical treatment. Clinical biomarkers may still present certain operator-dependent and time-demanding disadvantages, which may be solved in the future by cumulating experience and automatisation of the procedures. At last, biomarkers are already applied in the longitudinal assessment of the disease course in patients included in HS registries, offering a better stratification of patients groups and evaluation of their therapeutic needs.

002 PL02 - oral session | Updated medical treatments of hidradenitis suppurativa, more options, less evidence

W. Gulliver

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Hidradenitis suppurativa (HS) is a chronic inflammatory skin disease characterized by painful recurrent nodules and abscesses that rupture and can lead to sinus tracts and scarring. Prior to 2016 we did not have an evidence-based therapeutic approach to HS. After publication of the European Guidelines on the Treatment of HS, a holistic, evidence-based approach which implemented Level of Evidence (LOE) and Strength of Recommendation (SOR) was published for the treatment of HS.

In 2018, many aspects of the evidence-based approach remain unchanged, including the diagnosis of HS being made by a dermatologist or other healthcare professional with expert knowledge in HS. As well all patients should be offered adjuvant therapy such as pain management, weight loss, tobacco cessation, treatment of infections and use of appropriate dressings. Treating physicians should be familiar with disease severity scores, especially Hurley Staging, HiSCR and the newly introduced scores including the IHS4 (International Hidradenitis Suppurativa Severity Score) and the SAHS (Severity Assessment of HS Score). Routine use of patient-reported outcomes using Visual Analog Scale (VAS) for itch and pain as well as DLQI, is recommended. Surgical intervention should be considered in all patients, depending upon the type of lesions and extent of scarring. An evidence-based surgical approach should be implemented.

When it comes to evidence-based medical therapy, we have reasonable evidence for first line therapies including Adalimumab (LOE Ia, SOR A), topical Clindamycin, oral tetracycline (LOE IIb, SOR B), or oral Clindamycin/Rifampicin (LOE III, SOR C). When it comes to second and third line therapies we have seen new studies with intralesional and systemic steroids, improving the evidence from LOE IV to III and SOR D to C. With respect to third line treatment, it is suggested that Isotretinoin be removed from the recommendations and Cyclosporine go from LOE of IV to III and SOR D to C.

In terms of new therapeutics recent studies suggest that Ustekinumab, Anakinra, Ertapenem, Apremilast, Secukinumab and Guselkumab be considered as third line treatment.

Even though the body of evidence we have to guide us in the treatment of HS continues to grow, most of the studies are based on case series and case reports, which in theory, gives the clinician many more options but still with most treatments having Level of Evidence III or IV and Strength of Recommendation C or D.

GENETICS

003 OS01-01 - oral session | Molecular profiling of hidradenitis suppurativa/acne inversa skin reveals immune related signatures influencing keratinocyte differentiation and innate immunity

C. C. Zouboulis[1,3]; A. Nogueira da Costa[2]; E. Makrantonaki[1]; A. M. Hossini[1]; X. X. Hou[1]; D. Almansouri[1]; N. Bonitsis[1]

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Hidradenitis suppurativa/acne inversa (HS) is a chronic inflammatory skin disease with recurrent nodules, abscesses, ans tunnels as well as scarring and suppuration of intertriginous areas. HS affects approximately 1 % of the population. It has been associated with smoking, obesity and increased risk of metabolic syndrome. Moreover, a variety of co-morbid disorders, such as inflammatory bowel diseases, spondylarthropathy and cardiovascular disorders may occur. However, the pathogenesis of the disease remains unclear.

To elucidate HS pathogenesis whole genome skin profiling was conducted to detect HS molecular taxonomy and provide insights on key signalling pathways responsible for the development of the disease. Experiments were performed in lesional and non-lesional skin of European Caucasian female HS patients (median age 34.5 years, range 27-53, n = 8) and matched healthy controls (median age 36.5 years, range 18-53, n = 8) using the Agilent array platform. Confirmatory quantitative reverse transcription polymerase chain reaction and targeted protein characterization was conducted via immunohistochemistry in an additional group of female patients (median age 42 years, range 38-48, n = 4). The study was approved by the Ethics Committee of the Charité Universitaetsmedizin Berlin.

The whole transcriptome comparison of lesional, non-lesional and healthy control skin revealed 13 genes to be HS characteristic from a molecular standpoint, providing evidence of potential key regulators of its pathology (DEFB4, MMP1, GJB2, PI3, KRT16, MMP9, SERPINB4, SERPINB3, SPRR3, S100A12, S100A7A, KRT6A, TMPRSS11D). The further validation at the mRNA and protein level supported the findings reported at the genome level. Brought together, these data propose alterations to key regulatory pathways involving glucocorticoid receptor signaling, atherosclerosis signaling, inhibition of matrix metalloproteases, HIF1α signaling and IL17A signalling. From a functional standpoint, cellular assembly, maintenance and movement, hematological system development and function, immune cell trafficking as well as antimicrobial response are key processes detected to be affected in HS. At last, from a cellular standpoint, epithelial and adipose tissues are involved.

Our data provides a characterization of HS molecular taxonomy and insights to the disease pathomechanisms. Furthermore, our findings highlight a panel of immune related drivers in HS, which influence innate immunity in keratinocytes and lipids producing cells.

004 OS01-02 - oral session | DNA methylation and microRNA biomarkers for noninvasive detection of hidradenitis suppurativa

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Hidradenitis suppurativa (HS) represent one of the most unpleasant and devastating skin diseases; patients with HS develop severe comorbidities and have an increased mortality risk. HS is often undiagnosed as there are currently no diagnostic tests. MicroRNAs (miRNAs) (21-22-nucleotide) are small noncoding RNAs have emerged as important epigenetic regulators that control gene expression in the human genome without altering the DNA sequence itself. MicroRNAs are being studied in different types of complex diseases including inflammatory diseases and various cancers, however there are not many studies have been done so far addressing the expression level of the microRNAs in relationship with HS.

In the current study, we measured methylation levels of miRNA encoding genes using the Illumina MethylationEPIC BeadChip array in blood DNAs of 24 HS subjects and an equal normal of controls (age and gender matched). Several "R" packages were used to identify the significant CpG targets of miRNA. We have performed bioinformatic analysis by mining the MicroRNA Target Prediction and Functional Study Database (miRDB) to predict the possible genes regulated by the identified methylated miRNAs.o

We have identified 49 differentially methylated miRNAs: including two important miRNAs, miR-142 and miR-1908. miR-142 was previously identified to be upregulated in atopic dermatitis patients (PMID: 27512348). miR-142 was also observed in infiltrated immune cells of psoriatic-involved epidermis (PMID: 24326350). We predict that, miR-1908 takes part in wound healing process as it targets SKI gene. SKI protein reduces scar formation by decreasing collagen production and relieving inflammation in rat. It is also observed in wounded skin of human tissues (PMID: 28289554).

This is the first study of the genome-wide methylation variation of global miRNA genes in HS subjects. The study presents a novel approach to understand the molecular mechanism of HS. Altered methylation of miRNAs detectable in blood may potentially be a sensitive biomarker to detect early risk of HS. Understanding the complex mechanism leads to the potential of using miRNAs as prognostic tools and therapeutic agents for HS.

005 OS01-03 - oral session | In familial cases hidradenitis suppurativa occurs earlier and more severely in children than in parents

A. Villani[1]; G. Damiani[2]; P. Pigatto[2]; E. Vilarrasa[3]; L. Puig[3]; J. Romani[4]; E. Agut-Busquet[4]; A. I. Liakou[5]; D. Rigopoulos[5]; D. Mintoff[6]; S. Aquilina[6]; T. Boye[7]; D. M. Saunte[8]; G. B. E. Jemec[8]; V. Del Marmol[9]; F. Benhadou[9]; D. Jullien[1]; P. Guillem[10]

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Although family history is reported by about one-third of patients with hidradenitis suppurativa (HS), little is known about potential intrafamilial variations. We therefore aimed to evaluate clinical phenotype in documented familial cases.

In this multicentre case-control study, we included parents and children from a same family, provided both were affected by HS and followed in the recruiting centre. Data were obtained regarding disease occurrence, severity, phenotype and comorbidities.

83 pairs (parent/child) were included from 9 centres. None of the child had both parents affected with HS. The sex ratio was not different between parents and children. Parents had a higher mean BMI (28 vs 26 kg/m², $P = 0.002$) and were more frequently smokers (87% vs 64%, $P < 0.001$). Disease occurred and was diagnosed earlier in children (16 vs 26 years, 22 vs 45 years, respectively, $P < 0.001$). Although not significantly, each body area evaluated (armpits, breast, buttocks, groins, pubis, genitals, anal region, neck) was less frequently affected and the mean number of affected areas was lower in children. Despite this, quality of life was more impaired in children (mean DLQI 14 vs 13, $P = 0.02$). Disease was more severe in children according to Hurley stage (I, II, III: 42% vs 39%, 28% vs 29%, 30% vs 33%, $P = 0.04$) and IHS4 (mild, moderate, severe: 31% vs 27%, 33% vs 27%, 36% vs 46%, $P = 0.136$). Comorbidities (joint disease, IBD, acne conglobata, pilonidal sinus disease and acne vulgaris) were similarly frequent between parents and children. Only 22% of the parents reported HS in their own parents, as compared to 100% of the children ($P < 0.001$). We hypothesized that HS may be worsened in children because of an additional inflammatory background transmitted by the parent non-affected with HS. None of the children had in fact such a context (inflammatory disease in the non-affected parent), while this was observed by contrast in 5% of the parents ($P = 0.125$).

It remains to be determined whether the differences observed in the study are related to the sole familial context (higher awareness of the disease, familial lifestyle induced by the parent's disease ...). An earlier occurrence of a genetically-determined disease is however suggestive of a mechanism called genetic anticipation. It is suspected when the symptoms of a genetic disorder are observed at an earlier age with each generation. An increase in disease severity is often associated. Genetic anticipation is common in inherited neurodegenerative disorders such as the Huntington's disease and the myotonic dystrophy. Other diseases, including inflammatory conditions (Crohn's disease, Behcet disease, rheumatoid arthritis, psoriasis ...) may also present genetic anticipation and HS could join this group. Mechanisms underlying our findings have to be identified, notably by exploring epigenetic and environmental factors.

006 OS01-04 - oral session | In familial hidradenitis suppurativa, the gender of the parent who transmits the disease influences children's phenotype

A. Villani[1]; G. Damiani[2]; P. Pigatto[2]; E. Vilarrasa[3]; L. Puig[3]; J. Romani[4]; E. Agut-Busquet[4]; A. I. Liakou[5]; D. Rigopoulos[5]; D. Mintoff[6]; S. Aquilina[6]; T. Boye[7]; D. M. Saunte[8]; G. B. E. Jemec[8]; V. Del Marmol[9]; F. Benhadou[9]; D. Jullien[1]; P. Guillem[10]

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A genetic background is supposed in hidradenitis suppurativa (HS) since a family history is found in about one third of the patients and mutations have been seldom identified in gamma-secretase genes. Only little data is however available about phenotype variations in familial cases.

In this multicentre case-control study (9 centres), we included patients with HS supposed to be transmitted from an affected parent. Patients were definitively included if HS diagnosis in the parent was

ascertained through direct clinical examination. Data were obtained regarding disease occurrence, severity, phenotype and comorbidities, depending on the gender of the affected parent.

We included 83 HS patients with a clinically ascertained diagnosis of HS in either the mother (n = 45, 54%) or the father (n = 38, 46%). There was no difference as regards the patients' gender, age at inclusion, BMI, smoking and quality of life evaluated by DLQI. Age at disease onset, age at diagnosis and diagnostic delay were higher when the affected parent was the father, although non-significantly (17 vs 16, 23 vs 20, and 6 vs 4 years, respectively). Patients with an affected mother were more frequently affected in the armpits (84% vs 74%, $P = 0.226$). By contrast, all other sites (breast, buttocks, groins, pubis, genitals, anal and neck) were more frequently affected in patients with an affected father. Differences were only significant for buttocks (55% vs 33%, $P = 0.045$) and genital area (45% vs 13%, $P = 0.001$). The mean number of affected sites was significantly higher when the affected parent was the father (4 vs 3, $P = 0.023$). As regards comorbidities (joint diseases, IBD, acne conglobata, pilonidal sinus disease, acne vulgaris, scalp folliculitis), there was also a trend for higher prevalence in patients with an affected father, the difference being significant for acne conglobata (29% vs 4%, $P = 0.002$) and acne vulgaris (58% vs 22%). There was no significant difference in IHS4 whether the affected parent was the father or the mother (mild/moderate/severe: 30%/22%/49% vs 25%/32%/43%, respectively, $P = 0.587$). However, disease severity appeared higher in patients with an affected father when evaluated through the Hurley score (I/II/III: 34%/18%/47% vs 42%/38%/20% if affected mother, $P = 0.021$).

The main limitation of this pilot study is the low number of patients. The results should therefore be confirmed by extended studies. Our results are however intriguing in that they suggest that the HS phenotype in familial cases could be influenced by the gender of the parent supposed to transmit the disease. It remains to be determined the underlying mechanisms: are they genetic or epigenetic (genomic imprinting) or environmental?

007 OS01-05 - oral session | MYH9: A newly identified mutation in 2 patients with steatocystoma multiplex suppurativa and their family members

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Steatocystoma multiplex suppurativa (SMS) is a rare skin condition characterized by the presence of multiple subcutaneous cysts and inflamed hidradenitis suppurativa (HS) like lesions. So far it remains unclear whether SMS is steatocystoma multiplex associated with HS, or if the inflammatory foci arise secondary from the existing cysts and thus mimic HS. Heterozygous mutations in the KRT17 (keratin 17) gene were described as the cause of the SMS. We report the first amutation in the MYH9 gene identified in 2 patients with SMS.

Two male patients (patient 1:45 years; patient 2: 37 years) with SMS introduced themselves in our outpatient HS consultation. Both presented with more than 100 non-inflammatory cysts spread over the whole body and several inflammatory foci, such as acute nodules or abscesses in the area of body folds. Patient 1 also suffered from psoriasis and reported severe hearing loss and the existence of lipomas in his mother and brother. Patient 2 reports similar cysts in his father.

Both patients underwent genetic testing, identifying a heterozygous mutation in the MYH9 gene (variant c.5695G>A; *P.* Glu 1899Lys) following an autosomal dominant inheritance. The mutation was also detected in the mother and brother of patient 1. Pathogenic changes in MYH9 have previously been associated with symptoms such as hearing loss, macrothrombocytopenia, nephropathy, cataracts and elevated liver enzymes with high intrafamilial variability. Consecutive tests confirmed a pathological audiogram and macrothrombocytopenia in patient 2.

We describe for the first time a mutation in the MYH9 gene in patients with SMS, which encodes a non-muscular myosin involved in cell motility and had previously been linked to a multisystem disorder affecting ears, eyes, thrombocytes, kidneys and liver with high intrafamilial variability. Further studies are needed to investigate the significance of this mutation in patients with SMS and perhaps cystic form of HS. Careful consideration should be given to possible comorbidities in patients with MYH9-associated SMS, such as hearing loss, thrombocytopenia, nephropathy elevated liver enzymes, as well as the possibility of transmission to their offspring in the care and counseling of our patients.

IMMUNOLOGY

008 OS02-01 - oral session | Hidradenitis suppurativa is an auto-inflammatory disease: results of an ex vivo study

A. R. J. V. Vossen; K. R. van Straalen; E. Florencia; E. P. Prens

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Immune dysregulation, carrying a strong IL-1 signature, is implicated in the pathogenesis of hidradenitis suppurativa (HS). Interleukin 1 production is based on activation of auto-inflammatory pathways including the inflammasome, and consequential induction of various cytokines and chemokines. The objective of this study was to determine whether the lesional HS cytokine protein and gene expression profiles could be mimicked/induced by stimulation of perilesional HS skin with IL-1α and IL-1ß.

Skin punch biopsies from normal appearing perilesional HS skin (HSP) were obtained from 10 HS patients and compared with

five skin samples from healthy controls (NN) . All samples were cultured for 24 h in a transwell system using a culture media and media containing either IL-1α 10 ng/mL or IL-1ß 10 ng/mL. Subsequently pro-inflammatory cytokine protein levels in the culture media using a customised 16-Plex Assay (Luminex) and mRNA expression of four pro-inflammatory markers in the skin biopsies using real-time quantitative PCR were analysed. First, the Mann-Whitney U test was used to compare protein concentrations in the culture media of HSP with NN skin. Second, the Wilcoxon signed rank test was used to pairwise compare stimulated conditions to a condition without stimulation, i.e. culture media (fold change = 1.00).

Overall 62.5% (10/16) of the inflammatory proteins were significantly elevated in HSP skin compared with NN skin. After stimulation with IL-1α or IL-1ß respectively 40.0% (6/15) and 78.6% (11/14) of the inflammatory proteins were significantly elevated in NN skin compared with 7.1% (1/14) and 38.5% (5/13) in HSP skin. Similar results were found for mRNA expression levels in stimulated HSP and NN skin. Altogether, cytokine levels in HSP skin are already upregulated and almost not further inducible by IL-1.

This study reveals the auto-inflammatory nature of HS, characterised by the spontaneous increased production of a broad range of pro- and anti-inflammatory cytokines. Our results show that IL-1ß is a more potent stimulus compared with IL-1α in both NN and HSP skin.

009 OS02-02 - oral session | The role of S100 proteins in hidradenitis suppurativa

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Hidradenitis suppurativa (HS) is a chronic, recurrent, debilitating inflammatory skin disease. The pathogenesis of HS is still largely unknown and requires further investigations. Recently, the important role of abnormal innate immune response in the pathogenesis of HS has been highlighted. The aberrant production of innate antimicrobial proteins has been suggested to play a crucial role in pathogenesis of HS. However, this issue is still not completely understood and further investigations are required. S100 proteins constitute the family of low molecular weight (9-13 kDa), calcium-binding proteins with some antimicrobial activity. In the last decades, S100 proteins have emerged as a key player of innate immunity, important in the pathogenesis of various chronic inflammatory disorders. Among S100 proteins, S100A7 (psoriasin), S100A8 (calgranulin A), S100A9 (cangranulin B), and S100A15 (koebnerisin) show antimicrobial activity and have been characterized as damage (danger) associated molecular patterns (DAMPs). In the presentation, we reviewed current knowledge and presented our own data concerning the potential role of S100 in the pathogenesis of HS and the significance of these proteins as a biomarker of disease activity.

Hidradenitis suppurativa (HS) is a chronic, recurrent, debilitating inflammatory skin disease. The pathogenesis of HS is still largely unknown and requires further investigations. Recently, the important role of abnormal innate immune response in the pathogenesis of HS has been highlighted. The aberrant production of innate antimicrobial proteins has been suggested to play a crucial role in pathogenesis of HS. However, this issue is still not completely understood and further investigations are required. S100 proteins constitute the family of low molecular weight (9-13 kDa), calcium-binding proteins with some antimicrobial activity. In the last decades, S100 proteins have emerged as a key player of innate immunity, important in the pathogenesis of various chronic inflammatory disorders. Among S100 proteins, S100A7 (psoriasin), S100A8 (calgranulin A), S100A9 (cangranulin B), and S100A15 (koebnerisin) show antimicrobial activity and have been characterized as damage (danger) associated molecular patterns (DAMPs). In the presentation, we reviewed current knowledge and presented our own data concerning the potential role of S100 in the pathogenesis of HS and the significance of these proteins as a biomarker of disease activity.

010 OS02-03 - oral session | The IL-1 pathway is hyperactive in hidradenitis suppurativa and promotes skin infiltration and destruction

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Hidradenitis suppurativa (HS; also designated as acne inversa) is a chronic inflammatory disease characterized by painful purulent skin lesions and progressive destruction of skin architecture. Despite the high burden for the patients, pathogenetic pathways underlying HS alterations remain obscure. Investigating the lesional HS cytokine pattern, IL-1b turned out as a highly prominent cytokine, being overexpressed even compared to psoriatic lesions. Analyses of IL-1b-induced transcriptome in various cell types disclosed an overlap of upregulated molecules causing immune cell infiltration and extracellular matrix degradation, as well as of specific cytokines including IL-6, IL-32, and IL-36. Matching cellular IL-1 receptor levels, dermal fibroblasts showed both the strongest and broadest IL-1b response, which was not clearly shared or strengthened by other cytokines. The IL-1b signature was specifically present in HS lesions and could be reversed by application of IL-1 receptor antagonist. In summary, the IL-1b pathway

011 OS02-04 - oral session | MAIT cells in hidradenitis suppurativa: friend or foe?

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Mucosal- associated invariant T (MAIT) cells are a novel subset of innate-like T cells. They are major players in the human immune system representing 2-10% of the human T-lymphocyte population. They are robust producers of effector cytokines such as IFN-γ, TNF-α and IL-17 an inflammatory mediator implicated in the pathogenesis of several inflammatory chronic diseases. Their presence in human skin has only recently been recognised with limited data in the literature and their involvement in hidradenitis suppurativa (HS) has not yet been explored. Our aims therefore were to characterize circulating and skin resident MAIT cell frequencies in HS patients and to assess MAIT cells for expressed molecules determining activation status, functionality and intracellular cytokine production.

Cross sectional prospective cohort study. Consecutive patients with a diagnosis of HS who attended the dermatology department were included. Following signed consent, clinical history and exam, a; blood sample and a 6 mm punch biopsy was taken from both involved and uninvolved skin.

Patients attending the dermatology department who did not have any inflammatory skin disease were also recruited as controls. CD45 positive cells were isolated from both the skin biopsies and peripheral blood samples. Enumeration and functional characterization of MAIT cells in the skin and blood was then performed by multi-colour intracellular flow cytometry. Immune parameters were correlated with disease severity index.

MAIT cell frequencies were reduced in peripheral blood samples of HS patients compared to age and sex matched controls (1.8% vs 4.5% in healthy controls). MAIT cell frequencies were increased in HS lesions when compared to matched blood samples and also compared to uninvolved skin (5% lesional skin vs 1.8% peripheral blood vs 1% uninvolved skin). Cytokine analysis demonstrated a significant increase in the frequencies of IL-17 producing MAIT cells in both the skin and blood of HS patients. To investigate the drivers of MAIT cell IL-17 production, we investigated cell-cell interactions and found IL-17 production by MAIT cells to be dependent on monocyte interactions.

We show that IL-17 + MAIT cells are enriched in the skin of HS patients. Furthermore, we show that IL-17 production by MAIT cells is dependent on interactions with monocytes. IL-17 is a pro- inflammatory cytokine which has been implicated in the pathogenesis of several chronic human diseases including in psoriasis. Collectively our data suggests that MAIT cells are contributing to the inflammatory milieu in HS, and their interactions with monocytes may represent a novel therapeutic target.

012 OS02-05 - oral session | Decreased serum level of interleukin-22 in patients with hidradenitis suppurativa

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Dysregulation of well-orchestrated pro-inflammatory cytokines interplay characterizes various chronic immune-mediated diseases. Current understanding of the underlying pathophysiology of hidradenitis suppurativa (HS) links the disease with proinflammatory activation and auto-immune processes. We investigated serum level of interleukin (IL)-22 - a cytokine critically involved in epithelial homeostasis – in a broad clinical spectrum of HS. Serum concentrations of IL-22 and IL-6 (as a marker of pro-inflammatory activation) were assessed in 74 patients with HS (37 men, age: 37 ± 10 years) and 15 healthy subjects. Clinical assessment of disease severity was made with Hurley staging.

Compared with healthy controls patients with HS demonstrated decreased level of serum IL-22 (16.8 ± 13.0 vs 33.8 ± 10.2 pg/mL, $P < 0.001$ vs controls). The mean level of IL-6 in the entire population was higher in comparison to controls but the difference did not reach statistical significance (5.88 ± 14.56 vs 1.18 ± 1.11 pg/mL, $P = 0.16$). Serum levels of IL-22 and IL-6 did not correlate with each other ($r = -0.17$, P =ns). We identified group of 24 HS patients with pro-inflammatory activation (as evidenced by elevated level of IL-6 ie > upper limit of normal in our laboratory) – in this group mean level of IL-22 was similar with the remaining patients (14.1 ± 10.9 vs 17.9 ± 14.4 pg/mL, P =ns). Disease severity (assessed with Hurley staging) did not differentiate IL-22 levels: 12.5 ± 5.5 vs 14.2 ± 10.8 vs 23.3 ± 18.0 pg/mL, Hurely I vs II vs III, respectively, $P = 0.13$).

Patients with HS demonstrate significantly decreased levels of serum IL-22 which was neither related to pro-inflammatory status nor associated with disease severity. The role of IL-22 in HS warrants further studies.

EPIDEMIOLOGY

013 OS03-01 - oral session | Hidradenitis suppurativa: disease burden on patients and the health care system in Germany - impact of severity and of duration of the disease

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The challenges in managing HS and the resulting disease burden for patients and the health care system have recently gained attention. According to the Canadian Hidradenitis Suppurativa Foundation, a patient has seen on average 5 doctors and had 17 appointments over 8 years until receiving the proper diagnosis.

The following research questions (RQ) have been defined to examine the burden of disease of HS patients in Germany:

1. Which therapies according to guidelines do patients receive and how high is the disease burden on the patient and the system?
2. Are there differences in disease burden of HS patients with respect to severity of the disease?
3. Is a longer existence of disease symptoms associated with higher severity?

The retrospective cross-sectional study was conducted by the evaluation of a questionnaire. Since April 2017, LAight® therapy for the treatment of HS/Ai in outpatient centers is offered in Germany. The analyzed data was collected in the period between October 2017 and March 2018 and refers to the situation of the last 24 months before the first-time treatment with LAight® therapy. All sites with membership in the DGfW (Deutsche Gesellschaft für Wundheilung und Wundbehandlung) offering LAight® were asked for participation in the study. This included 10 locations of the WZ®-WundZentren GmbH (in North Rhine-Westphalia, Baden Württemberg, Bavaria and Brandenburg) and my dermatologic office in Rhineland-Palatinate.

The data was collected partly through questionnaires, but also through the standard parameters entered in the documentation software when carrying out a LAight® therapy. Matching of the data was ensured by a patient-specific identifier.

In addition to key demographics, Hurley disease classification and number of sites involved, key risk factors (smoking behavior and BMI), quality of life (DLQI), and pain level (NRS) were recorded. Information on relevant limitations in professional and social life as well as applied treatment options recommended under the guidelines were also asked for. For RQ 1 we report adequate descriptive statistics, RQ 2 and 3 were tested by applying a one-way ANOVA.

The sample included 287 questionnaires matched with 242 data sets from the documentation software. As other studies showed, the impact of HS on professional and social life is high. The results of the ANOVAs on RQ 2 confirmed the assumption that disease burden and impact on the health care system significantly increase with Hurley stage and the results on RQ 3 showed a significant association of Hurley stage with duration of the disease ($P = 0.004$).

Data confirms that HS is a disease which carries a high disease burden increasing significantly with disease severity which in turn increases over time. Getting an early diagnosis and receiving effective treatment is crucial to prevent high personal and social costs.

014 OS03-02 - oral session | The Lithuanian prospective study of patients with hidradenitis suppurativa based on clinical outcomes proposed by the European Reference Network for Rare and Complex Skin Disorders (ERN-Skin)

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Hidradenitis suppurativa (HS) is affecting 0.4-4% of population. According to National Health Insurance fund's official information, the incidence of HS in Lithuania during 2006-2015 was 23.5 cases per 100 000 population with female to male ratio 2:1. The average age of patients was 33 years. Only 7.7% of patients have been hospitalized, while outpatient management was used in 92.3% of cases. Till 2016, there were not used standardized outcome measures for HS activity and therapeutic effects.

The demographic and clinical data of total 35 HS patients, included over 3 years period (2016-2018) into local database of the referent centre of Rare Skin Diseases in Kauno klinikos, were analysed.

Demographic results: of total, 51.4% were females. The mean age of patients was 38.6 years and the majority of them 68.6% (95% CI 52.02-81.45) live in urban area. Median age was 39 years when the disease was diagnosed and median duration of the diagnosis delay was 4 years. One of three patients (31.4% (95% CI 18.55-47.98)) had BMI ≥30 kg/m2 – obesity, 14.3% (95% CI 6.26-29.38) of patients had BMI 25-29.9 kg/m2 - overweight.

20% (95%CI 8.1-32.68) were of Hurley I stage, 45.7% (95% CI 30.46-61.81) of Hurley II and 17.1% (95%CI 8.1-32.68) of Hurley III stage. The median evaluation of PGA was 3 points and the average of DLQI was 10.9 before the treatment. Half of patients (48.6% (95% CI 32.99-64.43)) had involvement of 3 or more body sites, 28.6% (95% CI 16.33-45.05) and 22.9% (95% CI 12.07-39.02) had involvement of 2 and 1 site, accordingly. Bacterial sample from abscesses/fistules was taken from 62.9% patients. Negative culture results were found in 72.7% of samples. Bacteriological analysis showed group-C beta-hemolytic streptococci (n = 1), S. aureus (n = 5), P. mirabilis (n = 1), P. aeruginosa (n = 1).

Combined medical and surgery treatment was applied to 51.4% and monotherapy (medical or surgery) to 48.57% of patients during period of the disease. Local antibiotics, mainly clindamycin, were used for 77.1% (95% CI 60.98-87.93) and antiseptic agents for 54.3% (95% CI 38.19-69.54) of patients. More than half patients 60.0% (95% CI 43.57-74.45) were treated with systemic antibiotics, 17.1% (95% CI 8.1-32.68) were prescribed retinoids, 4 patients have started biological therapy with adalimumab in 2017 year but 1 patient has stopped after 6 months of treatment because of adverse event – pulmonary

tuberculosis. Surgical interventions were made to 51.4% of cases. Totally 40.0% of patients were hospitalized for treatment at least once. Standardized collection of epidemiological and clinical data helps to improve the quality of HS patients' healthcare. As agreed scales and forms are not always helpful to evaluate the severity of HS and treatment effect, the implemented standardized sonography could support the management of HS patients.

015 OS03-03 - oral session | European registry for hidradenitis suppurativa (ERHS): state of play 2018

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In 2015, the European Hidradenitis Suppurativa (HS) Foundation (EHSF e.V.) started developing a European Registry of HS (ERHS) in order to acquire further knowledge about the disease. The number of medical centers interested in the project has increased constantly, even beyond Europe. It is essential to keep the registry up to date by including recent innovations such as new treatments, clinical classifications, and scores. OpenClinica, a freely available software for electronic data capture and management was initially proposed to manage the information collected from patients. Unfortunately, some technical difficulties arose (e.g. installation), therefore alternative solutions might be employed in the future.

We record on a yearly basis the number of participating centers and their status of progress (installation and number of patients). During 2018, the two questionnaires of the registry (first visit and follow-up) were updated to include new treatments, clinical classifications, and scores. Regarding de the collection of the data, in our center (Erasme Hospital, Brussel, Belgium), we will abandon OpenClinica and start using the REDCAP software. The program writing of REDCAP is in progress in our center and we will provide the templates to anyone interested in using this software. For the participants who are not able to use the open source softwares, there is a new proposal of a central database, lead by Mathias Augustin (Germany), with the help of Vahid Djamei (IT engineer in charge of the project).

We obtained updates regarding: (a) the number of participating centers and their status of progress; (b) the two questionnaires of the registry (first visit and follow-up); and (c) the options to collect the data (Redcap and the Central database).

The ERHS is a flexible project which is also a promising tool for future understanding of HS. The most important is to use the common questionnaires to be able in the future to merge the different databases from different participating centers.

The project is supported by the European Academy of Dermatology and Venereology (EADV).

016 OS03-04 - oral session | Prevalence of Hidradenitis Suppurativa in German working population

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Hidradenitis suppurativa (HS) is a chronic inflammatory skin disease characterized by inflammatory lesions such as inflammatory nodules, abscesses and fistulas, associated with a severe impairment of quality of life. The data on prevalence vary greatly between the individual studies, so that the true prevalence is still unclear.

The aim of our study was to determine the prevalence of HS in the working population and possible comorbidities.

20112 employees in 343 German companies were dermatologically examined for HS-characteristics, the searched items were: abscesses single and confluent, painful nodules, skin fistulas and inflammatory papules. Patients with at least one defined characteristic were considered as prevalent. Point prevalence rates were calculated and differences between participants with and without Hidradenitis suppurativa traits were tested with Chi^2 tests.

From 20112, a total of n = 57 persons (0.3%) had HS-characteristics, of which 35 were men (61.4%). There was no significant difference by sex, age or skin type. Patients with HS compared to persons without showed significantly more frequent occurrence of at least one inflammatory skin disease and folliculitis.

In our cohort a prevalence of 0.3 % for HS specific characteristics could be determined. These prevalence values correspond to prevalences published in the literature. A limitation of our study is the voluntary nature of the study, which could lead to an underestimation of the true prevalence. It should also be noted that patients with advanced disease and acute relapses are often unable to work and are therefore not included in our cohort.

017 OS03-05 - oral session | A retrospective study of the characteristics of patients with early-onset hidradenitis suppurativa

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Hidradenitis suppurativa (HS) is classically described as a disease occurring within the third decade. Earlier occurrence is however not rarely observed in clinical practice but it has been poorly documented. The aim of the study was to evaluate the proportion of early occurrence (< 20 years) and the associated phenotype in a large cohort of HS patients; We included all HS patients from our centre for whom the age of onset of the disease was recorded.

We included 1197 patients (mean age: 35 years, female: 64%). The mean age at disease occurrence was 21 ± 8 years (IQR: 15-25). An early occurrence was observed in 627 patients (52%). Patients with an early occurrence were more frequently women (71% vs 57%, $P < 0.001$) and had more frequently a familial history of HS (28% vs 21%, $P = 0.005$). Although diagnosed as expected at a younger age, they had a longer diagnostic delay (9 vs 6 years, $P < 0.001$). Disease severity, as assessed by the Hurley score was not different in patients with an early occurrence. In these patients, the first sites affected by the disease were groins, (33 vs 30% in patients with a disease occurrence ≥ 20 years, NS), armpits (21 vs 27%, $P = 0.01$), buttocks (13 vs 7%, $P = 0.033$), gluteal fold (13 vs 12%, NS), neck (7 vs 5%, NS), intermammary fold (4 vs 1%, $P < 0.001$), inner tights (4 vs 5%, NS), pubis (2 vs 3%, NS), genitals (2 vs 2%, NS), breast (1 vs 2%, NS), anal area (1 vs 4%, $P < 0.001$), and others (3 vs 2%, NS). Patients with early occurrence developed lesions in groins in 78% (vs 72% in patients with a disease occurrence ≥ 20 years, $P = 0.011$), armpits (71% vs 67%, NS), buttocks in 40% (vs 28%, $P < 0.001$), gluteal fold in 37% (vs 34%, NS), breast in 25% (vs 15% in other patients, $P < 0.001$), inner tights in 22% (vs 17%, $P = 0.029$), pubis in 22% (vs 22%, NS), neck in 19% (vs 12%, $P = 0.004$), genitals in 18% (vs 24%, $P = 0.007$), intermammary fold in 11% (vs 4%, $P < 0.001$), anal area in 11% (vs 15%, $P = 0.036$), and other sites in 18% (vs 16%, NS). The prevalence of joint diseases, inflammatory bowel diseases and dissecting folliculitis of the scalp was independent from the disease occurrence. Acne vulgaris and acne conglobata were more frequent in patients with an early occurrence (20 vs 15%, $P = 0.016$ and 4 vs 2%, $P = 0.027$, respectively).

It should not be longer stated that HS is a third decade disease. Awareness of the possibility of an early occurrence is mandatory to help reduction in diagnostic delay. Although disease severity seems to be independent from occurrence, some sites are significantly more or less frequently affected in patients with an early occurrence. Underlying mechanisms have to be determined including genetics (disease occurs earlier in familial cases) as well as epigenetic and environmental factors.

MEDICAL TREATMENT 1

018 OS04-01 - oral session | Intralesional triamcinolone for fistulous tracts in hidradenitis suppurativa: an uncontrolled prospective trial with clinical and ultrasonographic follow-up

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The study objective was to assess the clinical and ultrasonographic (US) response to intralesional triamcinolone for fistulous tracts in Hidradenitis Suppurativa (HS).

A prospective open-label study was conducted to assess the clinical and US response to intralesional triamcinolone (40 mg/mL) for fistulous tracts in HS. Consecutive patients (Hurley II stage exclusively) attending to our department were offered participation from August 2016 to August 2018. A single intralesional triamcinolone (ILT) administration was the only treatment prescribed. Patients on systemic medications or those who were candidates for these drugs were not included in the study. A single fistulous tract was chosen per patient for treatment and follow-up. The lesion was chosen based on its size (less than 25 mm in length after US examination) and grade of inflammation. Each patient was evaluated at 2 times-points (at day 0 and day 90). The lesion was marked and a clinical picture was taken at every visit. Demographic and HS-related information were recorded. Clinical information such as, size, location, duration, pain, itch side effects, and clinical resolution were recorded. US findings, such as size, diameter of the fistulous tract, colour doppler activity and US resolution were also recorded.

Overall, 53 HS-patients with fistulous tracts were recruited. Out of the 53 patients, 36 (68%) were female. 30 (56.6%) of the patients were smokers and 36 (67.9%) were obese or overweight (BMI≥25 kg/m^2). Median Sartorius score was 9.0 (IQR 9.0-36.0), and median DLQI was 4.0 (IQR 1.0-10.0). Median duration of the lesion treated was 6 months (IQR 3.0-12.0). The most common affected site was the groin (45%). Mean clinical size of the lesions was 17.0 mm (SD) (5.1) while mean US size was smaller at 16.0 (4.9) mm. Positive color Doppler activity was found in 10 lesions (19.9%). Out of the 53 patients, 7 were lost to follow up after the baseline visit, therefore outcome measures included 46 patients. The fistulous tracts were injected with a mean of 0.49 mL triamcinolone 40 mg/mL(range 0.2-1.0 mL). 20 (43.5%) lesions showed clinical and US resolution at day 90. However, 13 (28.3%) showed only clinical resolution while persisting on the US, and 13 (28.3%) persisted both clinically and on US. At follow up visit, mean clinical size was reduced from 17.0 mm to 5.1 mm ($P < .0001$). US measurements also showed a significant reduction in mean length from 16.0 mm to 8.6 mm ($P < .0001$) and mean diameter from 2.7 mm to 1.4 mm ($P < .0001$). Significant improvement was also found in other outcome measures such erythema, edema, suppuration, pain and pruritus. Cutaneous adverse effects were common, appearing in 26 (56.6%) of patients. Pigmentation changes were the most commonly found side effect, present in 25 (54.3%).

Our study suggests that ILT is perceived as beneficial by physicians and patients in the management of small fistulous lesions in HS.

019 OS04-02 - oral session | Oral clindamycin and rifampicin in the treatment of hidradenitis suppurativa - acne inversa in paediatric age: a prospective study

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Hidradenitis suppurativa–acne inversa (HS-AI) is a chronic, inflammatory, recurrent, debilitating skin disease of the terminal hair follicle that usually presents with painful, deep-seated, inflamed lesions in the apocrine-gland-bearing areas of the body.

The disease typically occurs in the second and third decade of life but an early onset, in paediatric age (0-16 years), is reported in 2% to 8.1% of patients.

No therapeutic trials have been published in paediatric HS-AI, and treatment recommendations are based on case reports and extrapolation of therapies tested in adult patients.

In adults, several case have described the beneficial effect of combination therapy using clindamycin 300 mg b.i.d. with rifampicin 600 mg once daily or 300 mg b.i.d. up to 10 weeks. Although no RCT has been conducted, the results of the published case series are very consistent and support the use of the combination. None of the published series include any paediatric patients, but both drugs are used in children to treat other infections, suggesting that this may be a viable approach to paediatric HS as well.

The study aim was to assess the effectiveness and safety of a 10-week combination of oral clindamycin (600 mg daily) and rifampicin (600 mg daily) in the treatment of HS-AI in paediatric age and to detect potential risk factors for non-response.

20 patients with age ≤16 years and affected by HS-AI were enrolled in a prospective non comparative study. The parameters used to evaluate the efficacy of the treatment were as follows: (i) severity of the disease, assessed with the Sartorius score before (T0) and after (T1) treatment and (ii) the number of exacerbations during the treatment period compared with those occurring in the previous three months.

All patients enrolled completed the study. Both Sartorius score and the number of exacerbations showed a significant reduction after treatment: P = 0.0000885 for Sartorius score and P = 0.000987 for the number of exacerbations. Three out of 20 patients complained of gastric side-effects. Among all the considered epidemiological and clinical data, none was found to be related with a significantly higher risk of failure to respond to treatment.

The combination therapy of clindamycin and rifampicin for 10 weeks was found to be an effective and safe therapy option in the treatment of HS-AI in paediatric age.

020 OS04-03 - oral session | Antiandrogens as a therapeutic option for hidradenitis suppurativa/acne inversa

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Hormones, and particularly androgens, have been described to play an important role to the pathogenesis of hidradenitis suppurativa/acne inversa (HS/AI). The prevalence of HS in women is almost three times higher than in men. Despite this, similarly to acne, no hyperandrogenemia has been detected in most cases, leading to the assumption that hyperandrogenism (an end-organ sensitivity to normal levels of circulating androgens or in-situ production of more potent androgens) can be an etiologic factor for HS. Therefore, antiandrogen treatment modalities were often utilized to treat different stages of HS severity. Based on this observation, we conducted a bibliographic research on articles, which document antiandrogen treatment of HS/AI.

We conducted a research on PubMed using the keywords "antiandrogen", "hidradenitis suppurativa" or "acne inversa". All the therapeutic regimens yielded from the initial search were subsequently searched in combination with the key phrase "hidradenitis suppurativa". The results are discussed below.

After excluding the case reports and narrative reviews, we managed to acquire 1 randomised control trial, 1 case control study and 5 case series. The main antiandrogenic regimes were cyproterone acetate, finasteride and spironololactone. A wide variety of patients were included in the aforementioned studies, both adults and adolescents, males and females, with treatment duration of 1-16 months. Response assessments included Physician's Global Assessment for HS (HSPGA), inflammatory lesion count, fistula count, numeric pain scale, flare frequency, hormonal profiles and other factors. Antiandrogens, even as monotherapy, reduced inflammatory lesions after a 3 week to 16-month administration, with overall good tolerability.

The use of antiandrogens for HS can be used primarily for certain subpopulations and HS phenotypes, especially if other comorbidities (hyperandrogenism, acne tarda, polycystic ovarian syndrome) are present. More randomized control trials, especially in early stages of HS severity, are required to establish robust, cost-effective strategies as either an add-on or an alternative to antibiotic therapy.

021 OS04-04 - oral session | Apremilast for moderate hidradenitis suppurativa: results of a randomized controlled trial

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Effective anti-inflammatory treatments for hidradenitis suppurativa (HS) are limited. We evaluated the efficacy and short-term safety of apremilast in patients with moderate HS.

Twenty patients with moderate HS were randomised in a 3 : 1 ratio, to receive blinded treatment with apremilast 30 mg twice daily or placebo for 16 weeks. The primary outcome was the Hidradenitis Suppurativa Clinical Response (HiSCR) at week 16. Linear mixed effects modeling was used to assess secondary clinical outcomes between treatment groups, including the abscess and nodule (AN) count, NRS pain and itch, DLQI, and expression of inflammatory markers in HS lesional skin.

The HiSCR was met in 8 of 15 (53.3%) patients in the apremilast group and none of 5 patients (0%) in the placebo group ($P = 0.055$)

at week 16. Moreover, apremilast-treated patients showed a significantly lower AN count (mean difference -2.6; 95% confidence interval -6.0, -0.9; P = 0.011), NRS for pain (-2.7; -4.5, -0.9; *P* = 0.009) and itch (-2.8; -5.0, -0.6; *P* = 0.015) over 16 weeks compared with placebo-treated patients. There was no significant difference in the DLQI over time between the two treatment groups (-3.4; -9.0, 2.3; *P* = 0.230). In HS lesional skin at baseline, protein levels of S100A12 and IL-17A were significantly elevated compared with nonlesional skin (FDR<0.044). These pro-inflammatory markers showed a clear, but nonsignificant (*P* > 0.05), decrease in response to apremilast, on the protein and mRNA level, respectively. The most frequently reported adverse events in the apremilast-treated patients were mild-to-moderate headache and gastro-intestinal symptoms, which have not resulted in drop-outs.

Apremilast at a dose of 30 mg twice daily demonstrated clinically meaningful efficacy and was generally well tolerated in patients with moderate HS. Protein levels of S100A12 and IL-17A were significantly elevated in HS lesional skin at baseline, and showed a downward trend in response to apremilast.

022 OS04-05 - oral session | Addition of methotrexate to anti-TNF-a (infliximab or adalimumab) at induction treatment in severe HS: improvement in therapeutic response and longer drug survival

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Infliximab and adalimumab are both associated with anti-drug antibodies (ADA), which explain the frequent secondary resistance observed with these 2 drugs when compared to more recent biologics, in which ADA levels are very low.

In most countries, adalimumab (ADB) is the only granted biotherapy to treat resistant HS, and infliximab (IFX) is widely used off label in hospital practice. As in psoriasis, after an initial good response, occurrence of ADA is often responsible for secondary drug failure.

In rheumatologic inflammatory diseases, MTX is often added to anti-TNF to avoid this mechanism and improve the response. We therefore designed a study to try to extend the therapeutic response in HS, and investigated the effect of MTX addition to the anti-TNF-Ab versus anti-TNF alone. We present here the preliminary results for the 32 first patients included, and the trial is ongoing.

It's a randomized prospective study planned to involve 240 patients with 2 cohorts: anti-TNF-a alone (infliximab or adalimumab) versus anti-TNF + MTX (20 mg/wk). The patients are all in II (16 patients) or III Hurley stages (16), and fulfil the European guidelines for anti-TNF prescription: 18 women and 14 men, mean age 34 y old (21-42). 8 patients have an associated psoriasis and 2 a Crohn's disease. The clinical evaluation is made at M3, M6 and M9 with HiSCR as main evaluating score, with HS-PGA. The 2 anti-TNF and the corresponding ADAs are systematically dosed at the same time (residual levels only, Test Lisa Tracker®), and correlated to the clinical response.

In the anti-TNF alone cohort, HiSCR was reached in 16 patients / 32 (50 %) after 6 months. In the second cohort with MTX added, HiSCR was reached in 23 patients (75 %) at M6 (*P* < 0.01).

In the 1st cohort, mean IFX level was 3.2 mg/mL (1.6-3.9) at M3, 2.7 at M6. Mean ADB level was 5.05 mg/mL (2.08-5.9) at M3, 2.88 at M6.

In the 2nd cohort with MTX, mean ADB level was 5.8 mg/mL (4.6-7.8) at M3, 5, 7 at M6. Results at M9 are not yet available. ADA levels: 1st cohort: < 4 mg/mL in 23 patients (not detectable), > 4 (detectable) in 9 patients at M3; and > 4 in 16 patients at M6. 2nd cohort (plus MTX): < 4 µg/mL in 28 patients, > 4 in 4 patients at M3; > 4 in 7 patients at M6.

These very preliminary results are very promising and show (for the 1st 32 patients) that addition of MTX to anti-TNF-a in severe HS improves the therapeutic response, and that this improvement is correlated with higher levels of circulating IFX or ADB, and lower presence of ADA at M3 and M6. These data are consistent with what's already known for rheumatologic and inflammatory bowel diseases. The study in now ongoing to complete all the planned inclusions.

BURDEN

023 OS05-01 - oral session | High prevalence of psychological traumas in patients with hidradenitis suppurativa (HS)

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Psychological support has been proposed in our centre to HS patients since 2012. Returns from the psychologists unexpectedly included a clinical impression highly prevalent psychological traumas.

The aim of the study was to evaluate the prevalence of psychological traumas in HS patients and their relationships with disease phenotype.

The charts of 155 patients (female: 72%, mean age: 33 years, Hurley I/II/III: 43%/38%/19%) who psychological support during a hospital stay for HS surgery between 2012 and 2017 were retrospectively reviewed. The psychological traumas potentially reported spontaneously by the patients during self-description of their emotional lives were recorded.

Up to 128 patients (83%) spontaneously reported at least one psychological trauma (1 event: n = 36; 2 events: n = 35; 3 events: n = 26; ≥ 4 events, n = 31). At least one trauma occurred during childhood in 74 patients (48%), in adulthood in 53 (34%), in both periods in 48 (31%).

Spontaneously-reported childhood traumas included separation from a relative (21%), psychological violence (15%), conflict family environment (9%), emotional deprivation (8%), serious illness of a relative (8%), sexual abuse (7%, including incest: 4%), death of a relative (5%), association to another personal serious illness (3%), family secret (3%), or were voluntarily left unexplained by the patients (3%).

Spontaneously-reported adulthood traumas included separation from a relative (23%), death of a relative (22%), serious illness of a relative (15%), related to work (11%), psychological violence (10%), family conflict (8%), related to HS management (8%), association to another personal serious illness (7%), physical or verbal assault (6%), domestic violence (6%), conjugal infidelity (6%), sexual assault (2%), family secret (1%) or were voluntarily left unexplained by the patients (15%).

The number of psychological traumas reported was higher in females than in males (2.5 vs 1.7, $P = 0.027$), especially for those occurring in adulthood (1.5 vs 0.8, $P = 0.009$, no difference for childhood traumas).

Patients reporting at least one psychological trauma during childhood had a significantly younger age at disease onset: 19 vs 22 years ($P = 0.027$). Regression analyses showed that psychological traumas could be related to affected sites. As an example, the death of a relative during childhood was an independent predictor of genital lesions (present in 75% of patients with vs 28% of patients without this experience, $P = 0.017$). Similarly, a family secret during childhood was an independent predictor of anal lesions (60% vs 19%, $P = 0.028$). Disease severity was higher in patients who underwent psychological violence during adulthood (I/II/III: 20%/33%/47% vs 46%/39%/16%, $P = 0.017$).

This pilot study strongly suggests that psychological management of HS patients should not only address HS impact on psychological health but also evaluate how psychological events could impact disease occurrence and evolution.

024 OS05-02 - oral session | Indirect self-destructiveness in hidradenitis suppurativa patients

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Hidradenitis suppurativa (HS) is a long-term, chronic, often debilitating skin condition which negatively affects patients' mental health and quality of life. Indirect self-destructiveness refers to activities extended over time, in which the person is not aware of their long-term harmful effects. These can include addictions, risky behaviours, neglects, resignation, helplessness. As such behaviours can be an additional factor impeding the achievement of positive clinical effects in the treatment of HS patients, therefore the objective of this study was to identify and assess the indirect self-destructive behaviors in patients suffering from HS.

The study involved 100 patients diagnosed with HS, recruited between January 2017 and October 2018. The study group consists of 59 males and 41 females aged 18-59 (mean, 34 ± 12.2 years). Disease severity was distributed as follows, Hurley I - 27%, Hurley II - 61%, Hurley III - 12%. The Polish version of the Chronic Self-Destructiveness Scale (CS-DS) Kelley was used to examine the intensity of indirect self-destructiveness in the studied population. The scale included the following domains: Transgression and Risk (A1), Poor Health Maintenance (A2), Personal and Social Neglects (A3), Lack of Planfulness (A4) and Helplessness, Passiveness in the Face of Problems/Difficulties (A5). Control group consisted of one hundred well-matched healthy individuals.

Preliminary results of our study revealed that, among patients suffering from HS, there are numerous manifestations of indirect self-destructiveness. HS patients in comparison to healthy controls showed increased indirect self-destructiveness in the majority of its domains. The most frequent were addictions in the form of alcohol consumption, smoking, social and health neglect. The detailed results will be given at the conference.

Indirect self-destructiveness plays an important role in HS patients and should be considered in the holistic approach of this group of patients.

025 OS05-03 - oral session | Influence of proper diagnosis of HS on patients daily life

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The time between first symptoms and the proper diagnosis in Hidradenitis Suppurativa (HS) still takes up to nine years. Different new non-surgical treatment options became available recently. As of now we don't know to which intent the correct diagnosis changes patients behaviour during acute inflammation, their approach to topical wound treatment and general understanding of disease. Also the average cost of wound treatment posing a potential burden to health care remains unclear.

We present a cross-sectional study that was performed in cooperation with LENICURA GmbH by evaluating an online survey uploaded on the company's website. The questionnaire was open for seven days from 4th June 2018 to 11th June 2018 to be completed anonymously online, with no link to personal data.

Patients were asked about their personal management of disease. Focussing on the method how they treated acute abscesses and what kind of bandage material was used. Another Focus was of the Questionnaire was how much patients payed on average for their Wound Dressings. Moreover, the survey included questions on washing and disinfection behaviours as well as shaving routines.

The survey included 171 completed questionnaires. On average patients need 25 min per day to care for their lesions and spend 32

€ per month on bandage material (both numbers increasing with Hurley stage). 61% of patients bought the material in the local pharmacy. Only 37% of material was being prescribed by a medical professional.

While 61% of special ointments are being mostly used in acute abscedation, self-employed incision or exprimation of abscesses still is performed in 45% of acute lesion. Only 16% see a doctor for an incision.

Even though considered a risk factor for exacerbation 70% of patients shave involved body sites and more than 80% of patients disinfect the involved sites at least once a day.

Data confirms that, even after proper Diagnosis, HS patients still take care of their disease mostly with over the counter products. Surprisingly few patients seek the help of a professional physician, rather than performing a self-employed incision of acute abscedation. Early diagnosis of disease remains to be one of the big challenges in treatment of HS. Patients still feel the need to take care of acute inflammation on their own. Pharmacies seem to be the most contacted institutions in daily HS management. Education of Pharmacists towards right wound care in HS might support a better medical supply to HS patients.

026 OS05-04 - oral session | Sexual dysfunction in patients with hidradenitis suppurativa: prevalence and risk factors

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Hidradenitis suppurativa (HS) is a chronic skin disease with high impact in quality of life. However, sexual health has scarcely been investigated in HS patients. The aims of this study are to describe the frequency of sexual dysfunction (SD) and to explore epidemiological and clinical factors potentially associated in patients with HS.

We conducted a cross-sectional study using a crowd-sourced online questionnaire hosted by the Spanish hidradenitis suppurativa patients' association (ASENDHI) from March 1st to April 1st 2018. Socio-demographic data, biometric parameters, medication and characteristics of the disease were collected. SD was assessed using Female Sexual Function Index-6 (FSFI-6) for women and International Index of Erectile Function-5 (IIEF-5) for men.

In total, 393 participants answered the questionnaire, 7 of them incompletely. The final sample consisted of 386 participants, 79.27% (306) women. The mean age was 37.81 ± 9.26. SD was found in 51% (95% CI 45%-57%) of women, and erectile dysfunction (ED) in 60% (95% CI 49%-70%) of men. Factors related to SD in women were education status (OR= 1.82, 95% CI 1,11-3; P <0.05), Patient's Global Assessment (PtGA) disease activity (β= -0.25 ± 0.11; P <0.05), Numeric Rating Scale (NRS) for pain (β= 0.1 ± 0.04; P <0.05) and unpleasant odour (β= 0.07 ± 0.03; P <0.05); being in a stable relationship was an important protector factor (OR=0.36, 95% CI 0.2-0,63; P <0.001). Factors related to ED in men were increasing age (β= 0.05 ± 0.02; P <0.05), the presence of active lesions in genital area (OR= 3.57, 95% CI 1.3-9.82; P <0.01) and the number of areas affected by active lesions (β= 0.32 ± 0.15; P <0.05).

We present the largest study regarding HS and SD. SD is very frequent among patients with HS. Potential risk factors of SD in women are education status, PtGA disease activity, NRS for pain and unpleasant odour and the absence of a stable relationship, and potential risk factors of ED in men are increasing age, the presence of active lesions in genital area and the number of areas affected by active lesions.

027 OS05-05 - oral session | Quality of life in hidradenitis suppurativa patients' partners: preliminary results

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Hidradenitis suppurativa (HS) is a chronic, debilitating disease leading to a patient's mutilation. It is also one of the most psychologically devastating diseases in dermatology. It is associated with chronic pain sensation and significantly affects quality of life (QoL). The objective of this study was to identify the QoL impairment in HS patient's partners and to assess its correlation with the severity of the disease.

A total of 50 pairs of HS patients and their partners were enrolled in this study. A detailed history and physical examination were performed. For assessment of HS severity, two standardized methods were used: the Hurley staging and Hidradenitis Suppurativa Severity Index (HSSI). The patients' partners QoL was measured using the validated Polish language version of Family Dermatology Life Quality Index (FDLQI).

Among the 50 HS patients 26 were men (52%) and 24 were women (48%). The mean FDLQI score for all patients was 8.7 ± 6.78 points, indicating a moderate effect on patients' partners' lives. The average FDLQI score in patients with Hurley I, II and III was 4.92 points, 9.4 points and 11.75 points, respectively. The most affected items of FDLQI that were reported by the patients' partners were: emotional distress, impact on the physical well-being, effect on leisure activities, some extra house-work and increased routine household expenditure.

A severe course of HS impairs the QoL of patients' partners, but its impact is still underestimated. Partners of HS patients suffer greatly from the emotional effects of living with, and caring for, a relative with a disease. They experience a negative influence on their family relationships, social lives, sex lives, leisure activities, hobbies, education, career and financial resources. Taking into account the burden of disease for the individuals and their partners can offer the clinicians a unique insight into issues, such as personal life and will lead to establishment of funding priorities and greater awareness of this condition.

OUTCOME MEASURES

028 OS06-01 - oral session | A composite biomarker score for the diagnosis of hidradenitis suppurativa

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One of the major drivers in the early diagnosis of hidradenitis suppurativa (HS) is the lack of diagnostic tools that can assist non-experts. This study aimed to the development of biomarker score for the diagnosis of HS.

Four cohorts of adults were studied: healthy volunteers (HV, n = 50), inflammatory bowel disease (IBD, n = 35), microbiologically confirmed staphylococcal acute bacterial skin and skin structure infections (ABSSSIs, n = 115) and HS (n = 196). Serum concentrations of interleukin (IL)-17, IL-20, IL-22, IL-23, cathelicidin (LL-37), s-E selectin, vascular endothelial growth factor-alpha (VEGF) and human-beta defensing-2 (HBD-2) were measured by enzyme immunosorbent assays. Following receiver operator characteristics curves and logistic regression analyses, a diagnostic algorithm was constructed.

Among the eight measured proteins, LL-37, s-E selectin, VEGF and hBD-2 at different thresholds were used at a three-step process. The first step excluded among all 396 studied individuals those being healthy. The second excluded the possibility among remaining patients those having IBD. At the last step, the algorithm considered diagnosis between HS and ABSSSIs and used s- E selectin (cut-off 60 ng/mL), VEGF (cut-off 1047 pg/mL) and hBD-2 (cut-off 1033 pg/mL) to develop a score. Values greater than 7 provided positive predictive value for HS 85.3%.

A diagnostic score for HS is developed based on different cut-offs of four measured serological biomarkers that can discriminate between HS and skin infections. The association of these protein biomarkers with skin innate immune responses reflects the clinical relevance of this score.

029 OS06-02 - oral session | The initial development of HiSQOL: a hidradenitis suppurativa specific quality of life instrument

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Hidradenitis Suppurativa (HS) is painful and HS lesions may, furthermore, be associated with pus and odour leading to significant stigma and negatively affecting the patients' health related quality of life (QOL). QOL is a multidimensional construct, which can be measured in various ways. It is generally acknowledged that generic or dermatologic QOL measures may not capture changes in QOL particularly affected in HS. Hence, the patients and experts included in the first HISTORIC (HIdradenitis SuppuraTiva cORe outcomes set International Collaboration) Delphi study reached consensus on including HS-specific QOL as a core domain to be measured in all future clinical HS trials. However, no validated HS specific QOL instrument exists to date.

To launch the development for a HS specific QOL instrument (HiSQOL, Hidradenitis Suppurativa Quality of life) was the main objective.

The initial phases of the questionnaire development, described here, included four major elements: item generation by patient interviews, development of a pilot questionnaire, questionnaire refinement and pilot testing.

For the item generation, 21 patients were interviewed individually or in focus groups. The interview analysis resulted in the identification of 105 candidate items. Next, questions were formulated representing each item with one or more different versions/formulations and the pilot questionnaire was developed. Finally, item reduction and two rounds of pilot testing resulted in a 23-item questionnaire. Both physical, psychological and social OQL dimensions were represented in the questionnaire.

In this study, we comprehensively explored the effects of HS on the QOL of the affected individuals and identified a 23-item HS specific QOL questionnaire. In the second round of pilot testing, the questionnaire proved to be feasible, acceptable and comprehensible. In future clinical HS trials, researchers will be able to measure HS-specific QOL with HiSQOL, potentially enabling them to discover more effective treatment options. We anticipate that a streamlined version of HISQOL may become available for clinical use in daily practice following thorough further validation of the questionnaire.

030 OS06-03 - oral session | QoL-HS - disease specific assessment of life in hidradenitis suppurativa

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Hidradenitis suppurativa is a chronic, debilitating skin disease with a high impact on quality of life. Assessment of disease related impairments in quality of life in patients with hidradenitis suppurativa (HS) requires specific tools. The aim of our project was to develop and validate a measuring instrument to assess disease specific quality of life instrument for patients with HS.

In the initial phase, n = 72 patients with HS were asked in an open text form about their burden of disease and treatment needs. We

collected the items and classified them in subgroups. The item pool thus generated was reviewed by a panel of dermatologists, psychologists and patients and transferred to a 26-item questionnaire. Each item was examined for its representativeness and its suitability with help of cognitive interviews. The validation study of the questionnaire is now ongoing in n = 100 patients. All patients have to complete the QoL-HS and two other QoL questionnaires (European Quality of Life-5 Dimensions, EQ-5D and Dermatology Life Quality Index (DLQI) at baseline and at the second time points (4 - 8 weeks).

We collected 34 different items for the question "the burden of disease". Top five of identified disease burden were: Pain (n = 41; 57%), restrictions in social life (n = 34; 47%), psychological burdens (n = 34; 47%), limitations in mobility (n = 30; 41%) and restrictions in spare time activities (n = 24; 33%). Top five of patient related treatment needs were reduction of pain (n = 51; 71%), no more limitations in mobility (n = 31; 43%), no more inflammation (n = 9; 12%), reduction of scars (n = 6; 8%) and weight loss (n = 3; 4%). We generated a questionnaire with 26 items. The validity is ongoing to prove internal consistency, convergent validity, feasibility and responsiveness.

QoI-HS is a new instrument to assess disease specific quality of life in patients with HS. The development of disease related measurement tools is an important step to improve the care of patients with HS.

031 OS06-04 - oral session | Superscoring hidradenitis suppurativa

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Hidradenitis suppurativa (HS) is a chronic, inflammatory, recurrent, debilitating skin disease. The scoring of HS is an important challenge: many scores exist, and all of them have their specificities, advantages and disadvantages.

This diversity of scores is problematic when carrying out clinical studies for example, because it is sometimes difficult to choose a score as "primary outcome".

We would like to present you a system of patients ranking, in order to match several scores. This way of combining multiple scores is thus what one might call a "superscore".

Our score will enable us to answer to two major questions: How to classify a cohort of patients from the most to the less severe case but also how to classify them from the best to the worst responder to a therapy based on the calculation of the existing clinical scores?

032 OS06-05 - oral session | Assessment of reliability of six commonly used scoring systems for hidradenitis suppurativa in a group of dermatology residents

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A wide variety of scoring systems have been proposed for assessing the severity of hidradenitis suppurativa (HS) until now. All of the available systems are based on the evaluation of clinical manifestations by a physician and in some cases an assessment of subjective symptoms given by the patient is also taken into account. None of the currently published assessment tools meets all the criteria required for an ideal score. The choice of the system depends on whether it is used for clinical trials or daily practice. The aim of our study was to compare and assess the reliability and reproducibility of 6 scoring systems commonly used for grading severity of HS: The Hurley staging, modified Hurley staging, Hidradenitis Suppurativa Severity Score System (IHS4), Hidradenitis Suppurativa Severity Index (HSSI), Sartorius Hidradenitis Suppurativa score and Hidradenitis Suppurativa Physician's Global Assessment Scale (HS-PGA).

On the scoring day, a group of 16 residents of dermatology were asked to evaluate 9 adult patients with HS with different severity of the disease using the Hurley staging, modified Hurley staging, IHS4, HSSI, Sartorius Hidradenitis Suppurativa Score and HS-PGA. All the subjects were assessed twice by each physician. Just before the scoring, all the assessors went through a short training about using the scoring tools and recording the data. Intra-rater reliability was assessed using intra-class correlation coefficient (ICC) and inter-rater variability was calculated using the coefficient of variation (CV).

In all 6 scorings the ICCs were > 0.75, indicating high intra-rater reliability of all presented scoring systems. The highest ICC was observed for the Sartorius Score, the Hurley staging and modified Hurley staging. The lowest ICC was noticed for the IHS4. On the other hand, the study has demonstrated moderate agreement between physicians in most of the presented scales. The CVs showed the highest inter-rater variability existing for the Sartorius Score. The lowest variability was found in assessing the Hurley staging, IHS4 and HSSI and these methods seem to be the most reproducible.

Comparing the Hurley staging, modified Hurley staging, IHS4, HSSI, Sartorius Score and HS-PGA, it was shown that ICCs were comparable, indicating high intra-rater reliability for all of the scales. None of the 6 evaluated scoring systems showed a significant advantage

over the other in that manner and none can be highly recommended as superior to the others. The inter-rater reliability occurs as much lower, and it was moderate for most of the assessments. However, the most repeatable results between researchers were obtained for the easiest scales. Hurley scoring, IHS4 and HSSI are only 3-graded scores which are quick and easy to perform, making it a suitable tool in everyday practice.

COMORBIDITIES

033 OS07-01 - oral session | Hidradenitis suppurativa comorbidities are similarly present in patients' relatives and influence patients' phenotype

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Several comorbidities have been described in association with hidradenitis suppurativa (HS). The aim of this study was to evaluate their prevalence in HS patients' relatives and their potential influence on patients' phenotype.

Since 2014, family history of HS, pilonidal sinus disease (PSD) and inflammatory or autoimmune diseases (INFL) were prospectively recorded in HS patients. Family history was considered positive whoever the affected relative (ascendants, siblings, descendants, or second degree relatives).

We included 1211 patients (female: 64%; mean age: 32 years; Hurley I/II/III: 58%/30%/12%). Family histories of HS (fHS), PSD (fPSD) and inflammatory diseases (fINFL) were observed in 303 (25%/first degree: 77%), 119 (10%/91%) and 121 patients (10%/82%), respectively. Patients with fINFL had significantly lower fHS (18% vs 26% if no fINFL, $P < 0.001$) while patients with fPSD had higher fHS (48% vs 23%, $P < 0.001$). No influence was observed between fINFL and fPSD.

Family inflammatory and autoimmune diseases included psoriasis (n = 45/4%), inflammatory bowel diseases (IBD, n = 27/2%), joint diseases (n = 21/2%), diabetes mellitus (n = 18/1%), lupus (n = 6), vitiligo (n = 5), multiple sclerosis (n = 5), scleroderma (n = 3), sarcoidosis (n = 2), Behcet (n = 1), Still (n = 1) and primary biliary cirrhosis (n = 1). All comorbidities were present with similar prevalence in patients and relatives, except joint diseases which were more significantly present in patients (n = 54%/4%) than relatives ($P < 0.001$).

Patients with fINFL were more frequently males (47% vs 35% if no fINFL, $P = 0.009$), while no difference was observed for neither fHS nor fPSD. While fHS was significantly associated with a younger age at disease onset (19 vs 21 years, $P < 0.001$), no difference was observed for neither fPSD (20 vs 21 years, $P = 0.067$) nor fINFL (21 vs 21 years, $P = 0.674$). Age at diagnosis, BMI, smoking and Hurley stage were not influenced by the presence of any family history. While fHS was associated with increased prevalence of IBD (4% vs 1%, $P = 0.003$) and acne vulgaris (23% vs 16%, $P = 0.008$) in HS patients, fPSD was associated with increased prevalence of PSD in patients (52% vs 34%, $P < 0.001$) and fINFL was associated with increased prevalence of joint diseases in patients (8% vs 4%, $P = 0.033$). If patients had a fHS, they were more frequently affected in the neck (13% vs 7%, $P = 0.004$) and the buttocks (38% vs 29%, $P = 0.002$) than patients with no fHS. Patients with fINFL were more frequently affected in the neck (16% vs 8%, $P = 0.005$) and the anal area (21% vs 12%, $P = 0.008$) than patients with no fINFL. No other site was influenced by the family histories.

Comorbidities usually associated with HS are also frequent in patients' relatives. In the context of known genetic background in HS, parents could therefore transmit HS predisposition not only if they have HS but also if they have a context of inflammatory or autoimmune disease.

034 OS07-02 - oral session | High prevalence of metabolic syndrome in patients with hidradenitis suppurativa, hospital-based study

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Metabolic syndrome (MetS), also known as syndrome X or dysmetabolic syndrome, refers to a cluster of metabolic conditions that can lead to heart disease, diabetes and other systemic diseases. Patient must have at least three of following five metabolic risk factors to be diagnosed with MetS – abdominal obesity, high triglyceride level, low HDL cholesterol level, hypertension and impaired fasting glucose (IFG). Hidradenitis suppurativa (HS) is a systemic disease which is considered to be associated with MetS. According to our experience, more than 50% of patients with HS suffer with one or more comorbidity. Diabetes, arterial hypertension, dyslipidemia and obesity are common. The aim of this study is to analyze the presence of risk factors for MetS in HS patients in the Bulgarian expert center for HS and to investigate whether MetS is more prevalent in comparison with the general population in Bulgaria

We analyzed data from 100 patients with HS, hospitalized in the Dermatology Clinic and Expert Center for HS at the University Hospital, Stara Zagora, Bulgaria. The patients were evaluated for overweight and obesity by Body Mass Index (BMI). Blood samples were collected for assessment of triglycerides, HDL cholesterol and fasting glucose levels. Blood pressure was also measured. Relevant medical history was recorded.

The prevalence of metabolic syndrome was 42% in the HS group compared to 26.8% in the general population. Three of the five metabolic risk factors were more prevalent in patients with HS – BMI

(HS - 54%, general population 37.5%), hypertriglyceridemia (HS - 35%, general population - 12.5%), impaired fasting glucose (HS - 36%, general population - 19%).

Metabolic syndrome is more common in HS patients than in healthy control population. Components of MetS were more prevalent in HS patients. As metabolic syndrome increases cardio-vascular risk, the factors comprising it should be considered in the comprehensive process of assessing and treating a patient with HS.

035 OS07-03 - oral session | Hidradenitis suppurativa and ulcerative colitis, difficult treatment to control both pathologies at the same time

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Hidradenitis suppurativa and Bowel disease (mainly Chron's disease) usually coexist in the same patient. Although both pathologies share ethiopatogenic mechanisms, they do not always respond in the same way to the treatments, especially biologic treatments.

We present a case of a 17 years old woman with ulcerative colitis and hidradenitis suppurativa, who is being treated with different treatments for both diseases since she was 13 years old (the age at which she was diagnosed). The problem with this patient is that even though different treatments have been tried for both pathologies, the responses have not always been always constantly fixed. Initially, she was treated with adalimumab (bowel disease dosage). The hidradenitis suppurativa achieved complete response, but ulcerative colitis did not response after more than 6 months of treatment.

Later on, adalimumab was changed to vedolizumab. Inflammatory bowel disease was controlled with that treatment, but skin disease worsened. Anticonceptives and antiandrogens were added, but hidradenitis suppurativa did not response. After one year with vedolizumab, bowel disease worsened, and the patient took some cycles of oral prednisone. When she was 16 years old, neither bowel disease nor skin disease was controlled by any treatment, so we decided to use off label ustekinumab for both diseases according to Chron's disease dosage. After 8 months with the treatment, bowel disease improved, with normal depositions and normal analytics parameters. Hidradenitis suppurativa was not controlled with that treatment, so 50 mg of sulfone were added. After 3 months with sulfone, cutaneous disease has been controlled, with only residual injuries. Now, both pathologies are controlled with ustekinumab with Chron's disease dosage plus sulfone 50 mg per day, and so the patient is able to lead a normal life. The patient is checked every 3 months as there could be risk that at any moment, either of both pathologies could be out of control again.

Adalimumab is a perfect treatment for hidradenitis suppurativa, but when the patient has comorbidities, we need to use other treatments. Sulfone is a good option without secondary effects.

036 OS07-04 - oral session | Hidradenitis suppurativa and psychiatric disorders: the Florence experience

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Hidradenitis suppurativa (HS) is a chronic inflammatory disease that primarily involves the skin, but which can be considered a systemic condition with multi-organ comorbidities including psychiatric and neurological disorders.

Indeed, skin, as the largest visible organ, plays an essential role in self-esteem and interpersonal relationships.

The aim of this study was to evaluate the prevalence of psychiatric and neurological disorders among HS patients. The study was conducted retrospectively among HS patients being treated at the Department of Dermatology of the University of Florence.

The study included 130 patients with HS: psychiatric disorders were diagnosed in 34 patients (26.1%), of which 28 females and 6 males. Depression was diagnosed in 20 patients (15.4%), intellectual disability in 10 patients (7.7%), feeding and eating disorders in 2 patients (1.5%), schizophrenia in 2 patients (1.5%). Anxiety (11.5%) coexists in 10 patients with depression, in patients with feeding and eating disorders and in 3 patients with intellectual disability.

From a neurological point of view 2 patients suffered from epilepsy.

In our experience anxiety and depression are the most common psychiatric comorbidities in patients with HS: this association, that goes beyond the body image disorder of HS patients, should be further investigated in order to take care of these patients in an integrated manner.

037 OS07-05 - oral session | The treatment of hidradenitis suppurativa associated with SAPHO syndrome

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SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) syndrome is a rare inflammatory condition that has a heterogeneous presentation of skin, bone and joints' combined manifestations.

We provide an up-to-date overview of current insight of the pathogenesis and different treatment options of SAPHO syndrome associated with hidradenitis suppurativa (HS). In addition, we introduce the case report on SAPHO syndrome which was recognized after severe painful attack of chest and shoulders after HS (Hurley stage II) treatment with clindamycin and rifampicin. Bone scintigraphy and CT findings supported the diagnosis of osteitis and arthritis of sternoclavicular joints. Medical history of the patient included low body

weight, depression and positive family history for acne. Initial oral treatment with isotretinoin showed worsening of acne conglobata. A good control of chest pain, acne, sterile pustulosis, and HS was achieved after short course of prednisone 20 mg/day and doxycycline 100 mg/day.

The etiology of SAPHO syndrome and HS is unknown. An initial bacterial trigger, mainly represented as cutaneous saprophyte Propionibacterium acnes, could take advantage of a selective deficiency of the innate immunity, implicating neutrophils. Nonsteroidal anti-inflammatory drugs, antibiotics, isotretinoin are not sufficient in such cases, and the use of tumour necrosis factor-α or IL-1 inhibitors may be necessary for permanent therapy. Our case shows that patient with HS should be screened carefully in clinical and radiological way for SAPHO syndrome. If both co-morbidities exist, better choice of treatment is immune system targeting therapy, but not antibiotics.

MEDICAL TREATMENT 2

038 OS08-01 - oral session | Intensification strategies 80 mg weekly with adalimumab in HS: when and what for?

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As Adalimumab is the only biologic treatment approved for the treatment of refractory severe hidradenitis suppurativa (HS) its use in alternative dosages depending on the patient status has not been addressed in literature.

We report our experience in three different clinical clinical scenarios in which Adalimumab 80 mg weekly was useful to improve disease control without switching to another off label biologic treatment or co-adjuvant treatment.

The clinical scenarios where 21 year old young patient with very limitating HS (IHS4 = 20) in whom 80 mg weekly was initiated in order to improve rapid time response (intensified start), a 48 year old patient in treatment with Adalimumab 40 mg weekly with apparent loss of response who improved with intensification adalimumab (80 mg weekly for 3 months) and later disintensification with stable disease and a patient with concomitant Crohn disease in treatment with Adalimumab 80 mg every two weeks who was intensified to 80 mg weekly for a severe Crohn and HS episode with excellent response

Intensification strategies of Adalimumab are useful in the management of patients with HS and should be considered as an alternative to switching or aggressive surgical or medical treatments.

039 OS08-02 - oral session | Efficacy of adalimumab in a patient with hidradenitis suppurativa and concomitant ulcerative colitis

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Inflammatory bowel disease (IBD) and hidradenitis suppurativa (HS) are both chronic inflammatory diseases with a big impact in the quality of life of the patients. It has been described a higher prevalence of HS among patients diagnosed with IBD, especially Crohn disease. Some drugs used in HS, such as antibiotics, can be harmful for IBD patients and they may trigger flare ups. Adalimumab is anti-TNF drug which is approved for the treatment of both diseases, being the doses similar in both disorders. We report a case of a patient suffering from HS and ulcerative colitis treated with adalimumab with excellent response in cutaneous and intestinal symptoms with a significant reduction in the International Hidradenitis Suppurativa Severity Score System (IHS4).

35-year-old male old diagnosed with ulcerative colitis of 9 years of evolution and hidradenitis suppurativa Hurley II-III and IHS4 20, of six years of evolution. The intestinal disease had been treated for a long time with mesalazine (oral and topical three times daily) and corticosteroids. He had received also multiple antibiotics for the treatment of his HS, with concurrent worsening of the inflammatory intestinal disease clinic. After ruling out latent infections such as tuberculosis, hepatitis and VIH, adalimumab was started by the dermatology department with an induction dose of 160 mg at week 1, with a maintaining dose of 80 mg every two weeks.

After three months of treatment the patient showed a spectacular response of his HS, with improve of the DLQI index and important reduction in IHS4, not having needed any antibiotic or drainage procedure during this time of treatment. Also the ulcerative colitis symptoms had diminished and the patient had not used any other treatment but topical mesalazine once a day and had the oral corticosteroids withdrawn. No adverse effects were reported.

Adalimumab is used in IBD and HS at similar doses and studies have proved the association of both disorders. We suggest considering this anti-TNF treatment for the patients with severe HS together with IBD. It is also important to have a multidisciplinary approach when managing these types of patients.

040 OS08-03 - oral session | Effectiveness of secukinumab (Anti-IL-17 Monoclonal Ab) in hidradenitis suppurativa: an open study (17 cases)

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The only approved biotherapy in Europe for HS id adalimumab. The effectiveness is not very High, as about 50% of the patient only reached the HiSCR in Pioneer studies. There is an urgent need for other systemic

or biological treatments. As for TNF-a, IL-17 levels are both elevated in tissue samples and blood in most patients. It seemed logical to try anti-IL-17 in HS, as they have shown also their effectiveness in psoriasis. Only isolated case reports have been published until now with good effect and we decided to pool the experience of 3 French centers involved in our national network (RESOVERNEUIL) in this open trial.

We included in an open study 17 patients, 10 women and 7 men, mean age 36.6 year-old (19-64). HS mean duration: 10.5 years (6-30). Hurley stages: respectively 5 Hurley III, 11 Hurley II, and 1 Hurley I. Mean weight 70 kgs (48-90). Secukinumab was initiated as for psoriasis: 300 mg weekly during a five-week period followed by 300 mg secukinumab monthly.

Comorbidities: 6 patient had concomitant psoriasis, 3 had inflammatory rheumatisms, one had PASH syndrome, 11 were still smokers (mean consumption 11 pack-years).

All patients were in therapeutic failure, after multiple treatments: all of them had received multiple antibiotics before, including ertapenem. All had received anti-TNF-a before (infliximab and/or adalimumab) during 4 to 24 months, and one even received anakinra during 2 years.

The chosen main efficacy endpoint was Hidradenitis Suppurativa Clinical Response (HiSCR, defined as ≥50% reduction in total abscess and inflammatory nodule [AN] count with no increase in abscess or draining fistula count). HS-PGA was also assessed.

13 out of the 17 patients (76%) had a favourable therapeutic response and reached the HiSCR. The HS-PGA rating fell from 4/5 to 2/5 in 16 weeks. One patient was lost to follow up.

In a follow up average of 14 months, we didn't observe any loss of response, even if flares were observed sometimes, but with return to the basic state.

The results didn't seem to be influenced by smoking status or weight in this little cohort.

An international phase III clinical trial is about to begin assaying secukinumab vs placebo. According to our very encouraging results (76% of overall response), such a trial is particularly interesting to conduct. Indeed, the only biological legally available, adalimumab, despite its official agreement, yields insufficient results (HiSCR in 50% of patients). In some published case reports, efficacy of secukinumab seemed to decrease over time. In our 13 responders, the clinical results appeared to be globally consistent over time (14 weeks in average), in spite of a few but flares. Indeed, blocking anti-IL-17 antibodies are very rare, reported in less than 1%. Our 3 centers will moreover participate in the future phase III clinical trial.

041 OS08-04 - oral session | Hidradenitis suppurativa successfully treated with ixekizumab – presentation of 2 cases

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Hidradenitis suppurativa is a chronic inflammatory skin disease that leads to a severe loss of quality of life. The disease is often intermittent and characterized by the development of inflammatory nodules, abscesses and draining fistulas. The exact pathogenesis are not yet fully understood, an increased expression of inflammatory cytokines such as IL-1ß, TNF-alpha, CXCL9 or IL-17A in lesional skin indicates an immunological genesis. First case reports on the efficacy of Secukinumab have been published. We report two cases of severe Hidradenitis suppurativa that were successfully treated with Ixekizumab.

Case report 1:

A 56 year old female patient presented with multiple diffuse fistulas and inflammatory nodules in the area of both groins and genital region (Hurley III, HS-PGA 5, ISH 55) in our Hidradenitis suppurativa consultation. After an antibiotic treatment with clindamycin and rifampicin 300 mg twice daily each, there was a slight improvement in fistula secretion. The patient reacted to the change to Adalimumab with a late type allergic reaction, so that we started a treatment with Guselkumab. As the therapy was not successful after 12 weeks, we switched to Ixekizumab. Already after 8 weeks a clear improvement of the findings was observed with only one active fistula (ISH 4).

Case report 2:

A 27-year-old female patient presented with mutilating inflammatory foci (fistulas and inflammatory nodules) in both groins and axillae after multiple previous surgeries (Hurley III, HS-PGA 5, ISH 35). After an initial improvement of inflammation under Adalimumab, a secondary loss of efficacy occurred about 9 months after the start of therapy with the occurence of a severely inflamed fistula in the area of the right groin to Mons pubis, so that the treatment was changed to Ixekizumab. After the reduction of the inflammatory reaction, the fistula was surgically removed. Of these, no new inflammatory lesions occurred 12 weeks after the start of therapy (ISH 0).

Therapy with Ixekizumab is a promising therapy option for patients with Hidradenitis suppurativa. Further studies on the larger patient population would be desirable to further investigate the efficacy in this indication.

042 OS08-05 - oral session | Guselkumab in the treatment of severe HS

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In this independent case series, we describe the efficacy of guselkumab, an anti IL-23 antibody in the treatment of severe hidradenitis suppurativa.

Beginning in 02/2018, three patients with severe hidradenitis suppurativa are treated with the IL-23 antibody guselkumab.

Patients were treated in the Department of Dermatology at the Klinikum Darmstadt GmbH; 3 patients with severe HS (Hurley stage III) who had responded poorly to other treatments or had any exclusion criteria for the use of adalimumab

Patients were treated with guselkumab according to the psoriasis dosing regimen. The treatment was assessed using the International

Hidradenitis Suppurativa Score System (IHS4), visual analog scale (VAS) for pain, and Dermatology Life Quality Index (DLQI) scores before therapy and 2 to 3 months after induction

All patients showed a significant improvement in IHS4 clinical staging. Moreover, patients reported a significant reduction regarding the VAS for pain and the DLQI.

SURGICAL TREATMENT

043 OS09-01 - oral session | Step by step surgery: a new therapeutic proposal for hidradenitis suppurativa

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Surgery represents a crucial therapeutic option for the management of Hurley II and III stages of hidradenitis suppurativa (HS), which feature scarring sequelae of the chronic inflammation that cannot be definitively cured by medical treatment. Radical surgery is indicated to effectively remove the disease: this means that large, destructing procedures have to be adopted for the most extensive cases, with heavy impact of patients' quality of life. Herein we propose a new surgical approach, the step-by-step surgery, aiming to reduce the post-surgical morbidity on patients by sequentially removing portions of diseased skin with secondary intention healing.

Patients with Hurley II and III HS were consecutively enrolled to receive step-by-step surgery either on the armpits or on the groin, under local anesthesia. Recurrence rate, as well as weekly-assessed patient-based scores [visual analogic scale (VAS) for pain, satisfaction, recommendation to other patients, and dermatology life quality index (DLQI)] were employed to assess the efficacy of the step-by-step surgery. Frequencies of contracture of the post-surgical scar and change of mobility of the shoulder joint were also collected.

A total of 12 patients with Hurley II and III HS received step-by-step surgery on the armpits (8/12) or on the groin (4/12). Recurrence rate was 1/12 (8.3%). Median VAS-pain, satisfaction, recommendation and DLQI were 7/10, 6/10, 6/10, and 15/30, respectively, at week 1; and 1/10, 9/10, 9.5/10, and 3/30, respectively. Contracture of the post-surgical scar was detected in 2/12 patients. Change of mobility of the shoulder joint was found in 8/8 patients who underwent axillary surgery, with improved mobility in 7/8 and decreased mobility in 1/8 patients.

We described a new surgical approach, the step-by-step surgery, which consists in consecutively removing portions of HS skin with secondary intention healing. This technique aims to reduce the post-surgical morbidity of patients (by eliminating the need of general anesthesia and skin grafts/flaps and by reducing the number of admission days and working days lost) and the costs related to the intervention. In this pilot study, the technique was well tolerated and produced very encouraging results.

044 OS09-02 - oral session | Wide surgical excision as treatment modality for hidradenitis suppurativa: a retrospective cohort study

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Hidradenitis suppurativa is a painful chronic, recurrent, debilitating inflammatory dermatoses characteristically involving inverse body parts like axillae, groins, anogenital and inframammary areas. Medical therapy is mainly aimed at reducing the incidence and duration of flare, but it is ineffective in preventing recurrence. Few published studies, mostly from developed countries, have reported use of wide surgical excision as a successful treatment modality for this condition and no study have been reported from Indian context. The current retrospective analysis was carried out to see the use of this surgical method in patients with HS and its success in preventing recurrence.

A Retrospective analysis for patients with Hurley's stage II (recurrent abscesses, single or multiple lesions with sinus tract and cicatrization) and stage III HS (diffuse and broad involvement with multiple interconnected sinus tracts) treated with wide local excision from the period of September, 2016 to June, 2018 at Department of Dermatology, Government Medical College and Hospital, Chandigarh, India was carried out. Wide local excision was performed under local anesthesia after taking proper consent. No graft was used post excision and wound allowed to heal by secondary intention. Patients with pregnancy or lactation, on anticoagulant drugs, coronary artery disease were excluded. Data collected consisted of patient's demographics, clinical characteristics, associated illness and previous and current treatments. Patients were followed from a period of 6 months to 2 years.

A total of 9 patients undergone wide local excision during the studied period. Mean age of the subjects was 28.66 ± 7.8 (SD)with mostly being in second and third decades of life. Male to Female ratio was 5:4. The mean duration of illness was 5.11 ± 3.48(SD) ranging from 1 to 12 years. Most of the subjects (77.8%) were either overweight or obese and in Hurley's stage II (77.8%). Axillae (88.8%) was most commonly involved followed by groin (33.3%) and buttocks (11.1%). No associated illnesses were seen in any of the cases. All patients were treated through serial wide excisions under local anesthesia followed by daily aseptic dressing and one week of systemic antibiotics. None of the patient showed recurrence of the lesions or any other complication associated with the procedure.

Wide surgical excision is definitive and safe modality of treatment in patients with severe Hidradenitis suppurativa and it results in long term disease control and improved quality of life.

045 OS09-03 - oral session | Systematic review of complications and recurrences following surgical interventions in hidradenitis suppurativa

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The possible connection between hidradenitis suppurativa (HS) patients undergoing surgery and higher complications/recurrences has been implied, but inconsistent results reported.

An objective was to create an overview and summary of complications and recurrences for HS patients undergoing surgery.

A systematic review was conducted by two reviewers. PubMed and Embase was searched using a predefined search string created in collaboration between the authors and a bibliographic fellow on 8th of December 2017.

Of the 230 references in the original search, 50 relevant articles were identified. This systematic review indicates an overall mean complication rate of 20.6% and a mean recurrence rate of 17.1% for HS patients undergoing surgery.

The heterogeneity and quality of the studies. Just one study met a grade A standard and a majority of studies were graded B. The studies uniformly lacked data of known comorbidities.

No significant association between these risk factors and surgical complications - or recurrence rates in this patient group was found. This review revealed a lack of quality and quantity data in studying the complications/recurrences in HS patients undergoing surgery, and it elucidates the need for better studies, and a necessity for a standardized definition of post-surgical HS recurrence.

046 OS09-04 - oral session | Rare forms and complex surgical situations in patients with hidradenitis suppurativa

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Both authors have extensive experience in the treatment of rare forms of hidradenitis suppurativa (HS) and complex surgical situations. Surgery is regarded as an important treatment option especially in moderate and severe cases. Surgery seems necessary, if irreversible tissue destruction is present, which is not suitable for medical approaches. A lot of cases can be sufficiently treated with radical excisions and postoperative wound care or if necessary reconstruction techniques.

However, especially in HS centers of excellence, we are often confronted with complex clinical findings and surgical pitfalls, like the following:

- Very extensive tissue involvement (deeply penetrating axillary area, contributing to arm contracture.
- Involvement of scrotal and gluteal areas - with perianal fistulas and deeper extensions/sinuses into the pelvis (perirectal/presacral/sacral bone).
- Secondary elephantiasis of external genital organs.
- Amyloidosis and cachexia.
- Development of neoplastic tumors, like: pseudoepitheliomatous hyperplasia, invasive squamous cell cancer (Marjolin's ulcer), or verrucous cancer.
- Coexisting Crohn's disease.
- Coexisting Darier's disease.
- Unusual location: on hairy skin (borderline cases with folliculitis decalvans of the scalp, on posterior neck (borderline with folliculitis keloidalis of the neck), of the male's chest – (borderline with acne conglobata of the back), of the female's chest, on coccygeal/sacral area (borderline with pilonidal cysts).
- HS with very early onset (children 13-14 years old).
- Wide-spreading HS with many disseminated small pathological structures (abscesses/cysts/ingrowing hair shafts) – (borderline with folliculitis).

The above mentioned situations and complex strategies will be presented and discussed.

NEWS FROM CLINICAL TRAILS

047 OS10-01 - oral session | IFX-1 in patients with moderate to severe hidradenitis suppurativa/acne inversa (HS): baseline characteristics of a double-blind, randomized phase 2b dose-finding study (SHINE)

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IFX-1, a novel monoclonal antibody blocking activated human complement factor 5a (C5a) has been shown to be safe and effective in an open label phase 2a study in patients with HS. A prospective, randomized, multicenter, double-blind, placebo-controlled phase 2b study in moderate to severe HS patients is currently ongoing in 9 countries (NCT03487276).

The primary objective is to evaluate a dose response signal of IFX-1 with Hidradenitis Suppurativa Clinical Response (HiSCR) at week 16. Adult patients with active, moderate to severe HS are enrolled and randomized into the study in either of four different dose regimens of IFX-1 or to placebo. The study was open for enrollment between February and October 2018. A snapshot of the clinical database was performed on 26 November 2018 and 169 patients were included in this analysis. The data are summarized by presenting mean ± standard deviation (SD) for continuous variables or the percentage of patients for categorical variables. As data cleaning is ongoing some variables are marked as missing.

Here, we report first blinded baseline data to characterize the study population. 56% of the patients were female and 44% male, 10% were black and 86% white. Mean ± SD age was 37.2 ± 11.4 years and weight 91.3 ± 18.0 kg. Median duration of HS was 8 years (range: 1 to 39 years). A family history of HS was documented in 24% of the patients. 60% and 39% of patients were classified as Hurley Stage II or III, respectively. The median inflammatory nodule count at baseline was 7 (range: 0 – 57), the median abscess count at baseline was 1 (range: 0 – 21) and the median draining fistula count at baseline was 2 (range: 0 – 20).

At baseline, demographic data and baseline characteristics according to the clinical severity are in line with the overall moderate to severe HS population, and similar to that of other clinical studies in this field.

048 OS10-02 - oral session | Long term results from a phase 2a clinical study with IFX-1 in severe hidradenitis suppurativa

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The efficacy and safety of IFX-1, a novel monoclonal antibody blocking complement split factor C5a, was evaluated in an open-label phase IIa clinical trial in 12 patients with severe hidradenitis suppurativa (HS). At the end of treatment period 75% of patients showed a response according to HS Clinical Response (HiSCR). After an 84-day follow-up period, 83% of the patients showed HiSCR response (NCT03001622). The objective of this retrospective long-term follow-up evaluation was to assess the long-term benefit of IFX-1 after cessation of the treatment.

A retrospective chart review was performed, and regular follow-up visits were recorded. Efficacy was assessed by AN count, number of draining fistula, and occurrence of flare-ups. Flare-ups were defined as deterioration of HS requiring antibiotic therapy.

Data from 10 out of 12 patients were available. In total 35 long-term-follow-up visits were documented (range 1 – 6 visits per patient). The median observation period from last IFX-1 treatment until end of long-term follow-up period was 226 days (range 119 – 324 days).

During the treatment period 2 flare-ups of HS occurred and two additional ones during the regular follow-up period in the Phase 2a study. In the long-term follow-up the number of flares was 26, ranging from 2 to 4 per patient. The median time to first flare after stop of IFX-1 treatment was 209 days (range 54 to 318 days).

The median AN count at baseline was 6 (range 3 – 11), at the end of the treatment period it was decreased to 2 (range 0 – 8) and at the last observation it was 4 (range 0 – 14). The median draining fistulas count decreased from 11 at baseline (range 3 – 21) to 9.5 at the end of the treatment period (range 0 – 8) and to 7.5 at the last observation visit (range 0 – 14).

Sustained remission was seen in most of the patients treated for 8 weeks with IFX-1. The median time to the first relapse was almost 7 months. These data support the further development of IFX-1 in HS.

049 OS10-03 - oral session | Efficacy of adalimumab in moderate to severe hidradenitis suppurativa: Real life data

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The hidradenitis suppurativa (HS) is a chronic, inflammatory skin disorder of the folliculopilosebaceous units. Even though there are therapeutic options, management of HS is challenging, since there are plenty of cases not responsive to treatment. Therefore, the approval of adalimumab for the treatment of moderate to severe HS enriched the therapeutic armamentarium and was proven to be promising for refractory HS cases. We describe our experience treating HS moderate-severe with adalimumab.

A single-center, observational, retrospective study was conducted to assess the efficacy of adalimumab on patients suffering from HS in daily practice. The records of HS patients who visited our department between January 2017 and October 2018 were used. Epidemiologic data, Hurley stage, affected locations, associated factors (obesity, tobacco), presence of other associated dermatological disorders, control of the disease and continuation or not with the treatment were recorded.

In total, 36 patients were included in the study. The ranged in age from 21 to 65 years old and included 16 men and 20 women, 69.4% were smokers and 61.1% presented overweight or obesity. Besides, 2 patients presented psoriasis, 2 Crohn disease and 2 acne. The most frequent location was armpits, followed by groin, inframammary and buttocks, although the majority presented several body areas affected at the same time or in the course of the disease. 44.4% had needed surgical intervention, the majority before to start the treatment. The average duration of treatment was 79 weeks (range 5-493). 72.2% were controlled and 4 patients was suspended due to inefficiency and 2 lost the monitoring.

This study provided evidence that adalimumab is an efficient and safe treatment option for moderate to severe HS. The patients tend

to have less frequent and intense outbreaks, although the majority often need the support of systemic antibiotic therapy. In addition to, after the start the treatment, the need to resort to surgery is less likely and it has a good security profile, in our series it hasn´t registered adverse effects that require to stop the treatment. Several biologic agents (infliximab, ustekinumab) have been used for the treatment of severe or recalcitrant to treatment HS. However, adalimumab is the only biologic agent that has been approved for the treatment of moderate to severe HS.

050 OS10-04 - oral session | The impact of hidradenitis suppurativa on absenteeism, reduced productivity, and work disability. Results from the PIRANHA study

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Debilitating chronic conditions and their treatments often negatively impact patient's ability to work, resulting in absenteeism and reduced productivity. The economic costs of this work impairment are immense. Hidradenitis suppurativa (HS) is a chronic inflammatory skin disease with a major impact on patient´s physical and psychological well being. Until today, there is little evidence on the impact of HS on patients' ability to work.

PIRANHA is a prospective non-interventional cohort study in HS. The data presented are based on the Work Ability Index (WAI) and the Work Productivity and Activity Impairment (WPAI) questionnaire for the subset of employed patients (interim analysis).

We analyzed the data of 416 patients included in the study. 54.1% of the patients were women, the median age was 39.7 (± 11.5) years, the BMI 31.0 (± 6.9) and the DLQI was 13.3 (± 8.1).

Analyzing patients´ occupation, 51.6% were employed full time (≥ 35 h/week), 19.7% at part-time (< 35 h/week), 7.0% were attending school/university/in training, 5.8% performed a domestic or educational work, 9.6% were unemployed and 6.4% retired. From the 22 retired patients, 5.3% were retired prematurely due to HS. In general, 4.5% of patients reported having changed their occupation due to HS. Patients reported 4.5 (± 3.7) h of sedentary work per day.

We found an overall low work ability (defined as a score <37.0 points on the WAI) of 28.8 (± 4.1). At the own work ability prognosis in 2 years (WAI dimension 6) 8.8% of patients find it unlikely and 26.6% are not certain to be able to work in two years from today. At the estimated work impairment due to disease (sub-score 4) 25.9% reported being entirely unable to work, 24.7% feeling able to do only part time work and 34.7% having to often or sometimes slow down their work pace or having to change their work methods. Only 9.9% of patients reported not being hindered at work by their disease.

41.9% of HS patients were unable to work because of HS within the last 6 months. Overall work impairment (productivity loss as measured by WPAI sub-score 3) was reported as 31.1% of the working capacity. Presenteeism (working while sick) caused 29.1% impairment while absenteeism caused 13.8% of working time being missed due to AI. Impairment of (non-work) activity impairment due to HS was of 39.8% (±32.0).

These results indicate that HS has a substantial impact on patients' work, causing absenteeism, impaired productivity, and work disability, stressing additional expenses of HS that are not related to medications or procedures.

051 OS10-05 - oral session | Impact of adalimumab on stabilization or sustained improvement in disease activity in moderate to severe hidradenitis suppurativa patients: an integrated analysis of PIONEER I and II trials through 36 weeks

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Hidradenitis Suppurativa (HS) is a painful, chronic, inflammatory skin disease characterized by recurrent inflamed nodules and abscesses; the clinical course varies and can progress from occasional solitary nodules to diffuse disease in multiple sites, and scarring. The Hidradenitis Suppurativa Clinical Response (HiSCR) includes a measure of worsening disease as defined by increases in abscess and fistula count. The objective of this analysis was to determine the impact of weekly adalimumab (ADA) on stabilization or sustained improvement of disease activity in patients with moderate-to-severe HS through 36 weeks of treatment.

In both trials, adult patients with moderate-to-severe HS for ≥1 year were randomized to ADA 40 mg weekly (ew) or placebo (pbo). This post hoc analysis included all patients receiving continuous ADA (pooled PIONEER I/II) or pbo (PIONEER II) through 36 weeks. Stability was defined as % of patients achieving no increase from baseline in abscess, fistulae or draining fistulae count for ≥3, ≥4, ≥5 consecutive visits. Sustained improvement was defined as % of patients achieving HiSCR or ≥25% improvement from baseline in abscess, fistulae or draining fistulae count for ≥3, ≥4, ≥5 consecutive visits. Median time to loss of disease activity stability or sustained improvement was assessed using Kaplan-Meier analyses. Descriptive statistics were calculated for all analyses.

99 ADAew patients and 151 pbo patients were included in this analysis. Lesion stability was achieved at ≥3/≥4/≥5 visits for ADA and pbo patients, respectively, for abscesses: 91.9%/86.9%/79.8% and 80.1%/65.6%/57.6%; fistulae: 83.8%/73.7%/66.7% and 72.8%/58.9%/53.6%; and draining fistulae: 90.9%/84.8%/75.8% and 70.2%/57.6%/51.0%.

Median time to loss of lesion stability (days) for ADA and pbo patients, respectively, for abscesses: not estimable (because <50% of patients lost stability) and 98

fistulae: 132 and 97; and draining fistulae: 183 and 79. Sustained improvement was achieved at ≥3/≥4/≥5 visits for ADA and pbo patients, respectively, for HiSCR: 57.6%/50.5%/42.4% and 28.5%/21.2%/16.6%; abscesses: 83.8%/77.8%/71.7% and 68.9%/61.6%/54.3%; fistulae: 46.5%/41.4%/39.4% and 41.7%/37.1%/30.5%; and draining fistulae: 81.8%/74.7%/63.6% and 56.3%/51.7%/48.3%.

Median time to loss of improvement (days) for ADA and pbo patients, respectively, for HiSCR: 58 and 17; abscesses: not estimable and 100; fistulae: 43 and 28; and draining fistulae: 127 and 83.

Through 36 weeks, a numerically greater proportion of ADA-treated patients experienced disease activity stabilization and sustained improvement across all measures; duration of stable disease activity or sustained improvement was longer with ADA treatment.

INNOVATIVE RESEARCH

052 OS11-01 - oral session | Deranged iron status in patients with hidradenitis suppurativa

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Iron deficiency is frequently present in inflammatory-mediated chronic diseases, irrespective of anaemia. Proinflammatory activation and auto-immune processes underlie pathophysiology of hidradenitis suppurativa (HS). Therefore, we hypothesize that in HS there are derangements in iron metabolism resulting in iron deficiency.

Serum concentrations of ferritin, transferrin saturation (Tsat), soluble transferrin receptor (sTfR), and hepcidin were assessed as the biomarkers of iron status in 74 patients with HS (37 men, age: 37 ± 10 years) and 44 healthy subjects. Iron deficiency was defined as ferritin <100 µg/L or ferritin 100-299 µg/L with Tsat <20% (following the definition used in the other studies in chronic disease). HS severity was assessed with Hidradenitis Suppurativa Severity Index and Hurley staging.

Compared with healthy controls patients with HS demonstrated deranged iron status as evidenced by decreased ferritin (91 ± 87 vs 157 ± 99 µg/L), Tsat (21.5 ± 10.8 vs 42.2 ± 11.7%) and low levels of hepcidin (a marker of depleted iron stores) (31.3 ± 25.9 vs 44.2 ± 22.0 ng/mL) (all $P < 0.05$ vs controls). There was also a trend toward higher values of sTfR (1.23 ± 0.35 vs 1.12 ± 0.19 mg/L) ($P = 0.09$ vs controls). In HS patients, none of iron status biomarkers correlated with the levels of interleukin-6 (pro-inflammatory activation). Additionally, disease severity did not differentiate iron status biomarkers. In HS patients iron deficiency was present in 75% and its prevalence was not related with disease severity (Hurely I/II/III – 82% vs 73% vs 67%).

Majority of HS patients demonstrate derangements in iron status typical for iron deficiency. These abnormalities are neither related to proinflammatory activation nor associated with disease severity. Whether it may have therapeutic impact needs to be further studied.

053 OS11-02 - oral session | Serum vitamin D in hidradenitis suppurativa patients: a pilot study

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Hidradenitis suppurativa (HS) is a chronic inflammatory skin disease which affects the follicles in apocrine sweat glands area of the skin, reaching 1–4% of the population. Recently, vitamin D deficiency has been associated with a number of immunoinflammatory dermatologic disorders, such as psoriasis, vitiligo, alopecia areata and lupus erythematosus.

The objective of our pilot study was to assess whether HS was associated with vitamin D deficiency and correlated with demographic and clinical characteristics of the patients.

The vitamin D level was determined in 22 HS patients followed at the Dermatology Unit of Florence University.

All 22 (100%) patients had vitamin deficiency (level < 30 ng/mL) which was severe in 19 (86%) patients (level < 20 ng/mL). In the logistic regression analysis, vitamin D deficiency was associated with HS independently of age, sex, and BMI. The vitamin D deficiency severity showed a mild correlation with the severity of the HS (r = -0.6) which signifies a possible link between the extent of vitamin D deficiency and the degree of the severity of HS, but it was not correlated with the disease duration and family history.

Our study confirms that vitamin D deficiency may be common in patients with HS and adds some important information concerning a possible link between this deficit and the severity of disease. Whether low vitamin D status is a modifiable risk factor in HS, we conclude that HS patients could be screened for vitamin D insufficiency for a more comprehensive management.

054 OS11-03 - oral session | Hidradenitis suppurativa in KID's syndrome: genetic characterisation, clinical and surgical intervention

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Syndromic hidradenitis suppurativa (HS) may occur in many syndromes, such as KID's syndrome (keratitis, ichthyosis and deafness). Mutations in the GBJ2 gene causes loss of function in connexin 26 protein, responsible for this syndrome. We report a case of KID's syndrome with a D50Y mutation and severe facial hidradenitis, recalcitrant to clinical treatments, who was submitted to a wide surgical excision. We discuss genetic and clinical aspects of HS in this rare disease, as well as the role of current and future biologics and the development of new targeted therapy.

The patient is a 14-year-old boy, the fifth concept after four miscarriages of a non consanguineous marriage. The patient was born premature, with diffuse skin rash. Deafness and photophobia were noticed since birth. At the age of 11 years, conglomerated papules and nodules occurred in the face and neck, as well as hyperkeratosis in hands and feet. Genomic DNA of peripheral blood was extracted using invitrogen protocol and standard Sanger sequencing directed to the hotspot area of GBJ2 exon gene, where we found the D50Y mutation. Oral isotretinoin was used (0,75 mg/kg/Day) for 6 months when the clinical picture resembled acne. Recurrent episodes of mucocutaneous candidiasis started with the use of oral antibiotics and Adalimumab was introduced, but with a low dose: 40 mg every other week (patient weight=35 kg), because of access problems, for 4 months. There was a rapid progression of the lesions and a wide surgical excision of the face using a skin graft from the legs was performed.

Genetic characterisation of the mutations in KID's syndrome with HS had been performed in about 7 cases (A40V, D50N, V27T and G12R) worldwide, but the D50Y mutation, already described in KID, is linked with these two conditions for the first time. Only one case (the D50N mutation) was treated successfully with Adalimumab. In our case, the first genetically identified KID's syndrome in Brazil, the patient failed biologic medication. This may have occurred because of inadequate dose of Adalimumab, a D50Y mutation (meaning that genotype may have a pharmacological correlation) or an advanced stage of the disease at the time of Adalimumab administration. Candidiasis infection had not worsened after anti-TNF treatment. This observation may reflect intrinsic TNF participation in immunodysregulation in this syndrome and also a non canonical pathway enrolled in Candida infection. As recent reports of secukinumab (anti IL17A) for HS are being published, careful attention must be taken with this specific population. Early treatment to prevent disfigurement must be instituted. In our case, the wide surgical excision was the option to stop progression. A novel human monoclonal antibody that modulates mutant connexin 26 protein is being designed and successful results in vitro were published recently. This may be a promising treatment for this syndrome

055 OS11-03 - oral session | Combined transcriptomic and immunohistochemical analysis of hidradenitis suppurativa lesions shows IL-17A pathway engagement

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Hidradenitis suppurativa (HS) is a chronic, debilitating disease with poorly understood pathophysiology where the unmet medical need for effective and safe treatments is high. Pathway knowledge outside TNFα and IL-1β is limited and an in-depth unbiased analysis of cell types and signaling cascades will help to dissect the underlying mechanisms and causes of the disease.

Due to the complex lesion topology in HS, a dataset combining transcriptomics with histological classification was generated. The aim of the study was to investigate pathway engagement of IL-17A and relevant cell types in HS lesions.

We analyzed lesional and non-lesional skin from surgical discards of patients with HS, Hurley stage II or III. Nineteen lesional and 12 non-lesional HS biopsies, as well as 8 biopsies from healthy volunteers were analyzed for global gene expression (Affymetrix microarrays). In addition, hematoxylin and eosin staining, and immunohistochemistry was performed on adjacent tissue.

Hierarchical clustering of the RNA transcript data grouped the HS and Healthy volunteers biopsies into lesional, peri-lesional and non-lesional/healthy groups, confirmed by histology. The global transcriptomics dataset was also analyzed for cell type-specific transcript signatures and IL-17 pathway signature using published and in-house signatures. Cell types identified in lesions included neutrophils, macrophages, B-cells, and various T-cell subsets like Th1 and Th17 cells. IL-17A signaling signatures derived from various cell types (keratinocytes, fibroblasts, whole skin) showed a significant increase in lesional samples, suggesting IL-17A pathway engagement. In addition, unbiased analysis of differentially expressed genes between the groups identified the canonical IL-17A pathway and various members of the IL-17 ligand/receptor family as upstream regulators.

Histopathology revealed severe mixed inflammatory infiltrates of mainly macrophages neutrophils, T and B cells as well as plasma cells in the lesional biopsies, thus confirming the transcriptional data. Immunohistochemistry showed expression of IL-17 in T cells and neutrophils. In addition, β-defensin 2 and S100A7A were both found in areas mirroring psoriasiform epidermal hyperplasia.

HS lesions show clear IL-17A pathway engagements on several levels. Inhibition of IL-17A signaling could be beneficial for HS patients.

056 OS11-04 - oral session | Cutaneous manifestations of the STAT1 mutation

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We present the case of a 29-year-old patient who suffered from chronic mucocutaneous candidiasis, recurrent respiratory infections, severe hidradenitis suppurativa, canker sores, lobular panniculitis without vasculitis, as well as early ovarian failure, growth hormone deficiency and congenital hypercalciuria.

The patient was diagnosed with a pathogenic mutation in STAT1 (transcription factor and signal transducer). These patients develop the main phenotype of chronic mucocutaneous candidiasis (CMC), and suffer other clinical manifestations such as increased sensitivity to Staphylococcus aureus infections, viral infections and autoimmune diseases.

STAT1 is a transcription factor that mediates the signaling of IFN alpha, beta and gamma. It is believed that exists an excessive response to inhibitory cytokines that inhibit development of IL-17 producing T cells (such as IFN alpha and gamma) and this fact increases the response to IL-6, IL-21 and IL-23 that favor the development of IL-17 producing T cells.

Regarding hidradenitis suppurativa (HS) there is a shortage of information on the pathogenesis and underlying immune dysregulation but it has been shown, that in the skin affected by lesions, there is an imbalance between Th17 cells and Tregulators cells being the first in greater proportion.

More studies are required to determine the possible dual role of immunity mediated by IL 17 since, as in our case, an excess of STAT1 activity is associated with CMC and autoimmunity

IMAGING

057 OS12-01 - oral session | Advances on ultrasound imaging of hidradenitis suppurativa

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The usage of ultrasound imaging in hidradenitis suppurativa (HS) has been growing in the last decade, and nowadays this imaging technique is widely used in several centers around the globe for supporting the management and research on HS.

In this session, we will review the last advances on ultrasound imaging of HS and the potential meaning of these advances. Images and videos extracted from the ultrasonographic examinations of HS patients that were performed with state-of-the-art ultrasound equipment, a clinical-sonographic correlation and a discussion of the findings under a physiopathological perspective will take place.

After completing this activity, the participant should be able to know the last advances on ultrasound imaging of HS and to correlate clinical and ultrasonographic findings in HS.

058 OSOS12-02 - oral session | Hidradenitis suppurativa phenotypes based on clinical and ultrasound characteristics

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Recently, the concept of window of opportunity in Hidradenitis Suppurativa (HS) patients has been proposed as the period during which efforts to control inflammatory activity may be most useful, that occurs in the early stages of the disease before the onset of sequelae and established irreversible damage. Immunomodulatory therapy during this period may alter the natural history of the disease by reducing the accumulation of tissue damage. However, well-defined phenotypes that may help in early detect those more aggressive patients are still lacking.

The objective of the present study was to define different profiles of HS patients based on clinical-ultrasound elementary lesions.

An observational, descriptive, nonrandomized, unicentric and retrospective study was conducted. Patients diagnosed with Hidradenitis Suppurativa in a specialized unit from May 2012 to April 2018 were included. Ultrasound test was used to better define all the elementary lesions that were present under the skin.

197 patients, 100 women and 97 men, aged between 25 and 47 years were included. Two well-defined subtypes, namely inflammatory and follicular variant, were defined. Whereas in the inflammatory subtype the mean age of onset was 26.69 ± 9.05, the mean age of onset in the follicular subtype was 17.62 ± 6.42. More nodules appeared in the follicular subtype (5.65 ± 3.38 versus 0.89 ± 2.72), while more abscesses and fistulas appeared in the inflammatory subtype (respectively 4 ± 2.74 and 3.11 ± 2.56 versus 0.56 ± 1.02 and 0.26 ± 0.56). IgA levels were significantly higher in the inflammatory subtype (497.71 ± 262.26 versus 232.38 ± 84.06). The occurrence of papular/nodular acne was higher in the follicular type (OR 3.167 [CI: 1.416-7.65]) and the conglobata/fulminans acne was higher in the inflammatory type (OR 0.075 [95% CI 0.02-0.222]). Pruritus was a more frequent symptom in the follicular type (OR 16.481 [CI: 8.225-34.479]) and odor and pain were more frequent in the inflammatory type (respectively OR 0.017 [CI:0.007-0.038] and (OR 0.063 [CI: 0.031-0.121]). In the follicular subtype the majority of the patients (70.14%) had PGA 3, while in the inflammatory subtype the majority (58.49%) had PGA 5. The mean HS4 was higher in the inflammatory subtype (21.04 ± 11.9) than in the follicular subtype (7.54 ± 4.66). The same applies to DLQI (15.9 ± 4.04, versus 9.78 ± 2.88). Regarding

location of the lesions, the groins were affected mostly in the inflammatory subtype, while the thighs were affected mostly in the follicular subtype. Finally, the inflammatory type was shown to be an independent risk factor of greater severity (OR 0.034 [95% CI 0.015-0.072]).
Two well-defined subtypes of HS, namely follicular and inflammatory phenotype, were characterized. Preliminary data suggest that the inflammatory variant is associated with a higher aggressiveness and a major risk of disease progression.

059 OS12-03 - oral session | Color doppler sonographic and epidemiological features of healing fistulous tracts in HS patients treated with systemic and biological treatments

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Hidradenitis suppurativa (HS) is a chronic inflammatory illness which is featured by recurring inflammation of hair follicles in apocrine areas. Progressive damage of follicular units leads to multiple fistulous tracts formation which are hallmark HS lesions and indicators of disease severity.
Therapy in HS is directed to reduce inflammation in HS lesions improving quality of life in these patients. However sonographic evidence of fistulous tract healing has not been addressed in the literature as a target of systemic or biologic therapies.
We reviewed our sonographic image database of 97 patients in follow up with the diagnosis of moderate and severe HS treated with systemic and biologic therapies in order to identify sonographic and epidemiologic predictors of complete sonographic fistulous tracts regression.
All examinations were performed with multichannel colour Doppler ultrasound equipment (My Lab Class C, Esaote, Genova) using variable frequency probes with an upper range up to 18 MHz and by the same dermatologist with more than 7 years of experience in dermatologic ultrasound. The fistulous tracts were examined in at least 2 perpendicular axes for assessing their long and short axes. Grayscale and color Doppler examinations with spectral curves analysis (vascularity) were carried out in all cases.
Epidemiological variables included age, sex, fistulous tract location, patient IHS4 score and fistulous tract remission time.
Measured sonographic variables were maximum fistula length, location (dermal subdermal or subcutaneous), presence or absence of fibrosis and subcutaneous edema (defined as hypoechogenicity between fatty lobules). Sonographic fistula type according Wortsman et al. classification was also evaluated by two dermatologist blind to patient's exploration and history.
Sonographic regression or healing of fistulous tracts was defined as minimal dermal sonographic changes after treatment with absence of Doppler flow at least 12 months of follow up; We found sonographic evidence of fistulous tract regression in 10 patients (5 = Adalimumab, 4 = antibiotics, 1 = secukinumab) who presented low or moderate inflammatory load (mean IHS4 = 8±4 mm) This fact indicates that these patient have a low inflammatory load most of the had an early response in the first 8 weeks of treatment.
Sonographic features of healing fistulae corresponded to middle size (mean 12 mm ±4 mm) type 2 fistulae (subdermal hypervascularized with minimum subcutaneous involvement) according Wortsman classification.
Fistulous tracts healing with systemic and biologic treatments in possible in patients with early middle size fistulous tracts with scarce subcutaneous inflammation with low inflammatory load, therefore initiation of systemic or biologic treatment in this subgroup of patients may avoid disease progression.

060 OS12-04 - oral session | Teleultrasonography for the early diagnosis of HS in the primary cares setting

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Teleultrasonography is a new telemedicine modality not previously consider for diagnosis in both dermatology or hidradenitis suppurativa (HS).
6 primary care centres connected with a tertiary care centre were consulted through the teledermatology system including ultrasonographic scan of the patients acquired by primary care providers. Images were analyzed by expert dermatologic ultrasound physicians and patients were appointed for in person confirmation.
A total of 94 tele ultrasound consultations were required by the primary care centres.
Correlation in general cases reached 92% while in the case of the 10 patients with HS, all patients were coincident for remote diagnoses.
Teleultrasonography is a useful tool for early diagnosis of HS in the primary care setting in collaboration with dermatologic ultrasound trained physicians

061 OS12-05 - oral session | Long-wave medical infrared thermography: a clinical biomarker of inflammation in hidradenitis suppurativa/acne inversa

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A more reliable classification of skin inflammation and severity of active disease results by ultrasound sonography and the new HS

classification system IHS4. However, an objective assessment of skin inflammation in a continuous mode is still the ultimate goal. Long-wave Medical Infrared Thermography (MIT) may offer a blood flow and temperature differential assessment in inflammatory conditions.

To evaluate the application of MIT in HS standardised photography of the areas involved or been candidate for HS involvement and MIT pictures of 18 patients [11 female, 7 male, median age 38.75 years (95% confidence intervals 28.5 and 51 years), Hurley score I 5.6%, Hurley score II 38.9% and Hurley score III 55.5%] were taken subsequently with a FLIR T650sc Thermography Imaging Unit at a distance of 50 cm from the skin surface and the pictures were superimposed. A modification of the Otsu's method facilitated the automatic lesion segmentation from the background, depicting the inflammation area. Moreover, MIT was administered in real-time mode during radical HS surgery.

A 1°C temperature difference from a corresponding symmetric body region was indicative of inflammation. MIT figures detected a gradual increase of skin temperature from 33.0°C in healthy skin in average to 35.0-36.6°C at the center of inflammatory lesions in the axilla and to 35.4-36.9°C at the center of inflammation at the groin area. Real-time MIT assessment enabled the definition of the margins and depth of the surgical intervention during the procedure.

MIT is a promising tool for detection of inflammation severity in HS lesions and can be used as a clinical biomarker in evaluation studies of medical and surgical HS treatment.

POSTER SESSION

062 PS01 - poster session | γ-secretase mutation in etiology of acne inversa

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Acne inversa (hidradenitis suppurativa) is a potentially severe and chronic inflammatory disease with a significant negative influence on the quality of life. Usually, lesions are located in the areas of skin folds and it is characterised by the presence of painful nodules and fistulas with a tendency to tissue fibrosis.

Currently, it is suggested that follicular occlusion by infundibular hyperkeratosis plays a crucial role in pathogenesis and an occupation of apocrine sweat glands is a secondary phenomenon. Most often, it refers to men after puberty.

Etiology of the inverted acne, as mentioned earlier, is not entirely clear. Familial occurrence of this disease was confirmed, but both genetics and inheritance pattern in most patients require further researches. In Chinese, Japanese and European families, the mutation of γ-secretase- NCSTN, PSENEN and PSEN1 genes was described. γ-secretase is one of many subunits of protease complex, which cleaves transmembrane proteins. It is also essential for an appropriate processing of the Notch protein, which is suggested as a key for the etiology of acne inversa. The best known substrate of γ-secretase is the amyloid precursor protein, which is an integral membrane protein. Thus, amyloid deposits were also described in patients with inverted acne.

063 PS02 - poster session | Hidradenitis suppurativa/Dowling-Degos disease (DDD) phenotype associated with mutations in NCSTN gene

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To assess the possible association between NCSTN gene mutation and Hidradenitis suppurativa (HS)/DDD phenotype patients. Mutations in this gene have been previously reported in HS.

Venous blood samples were obtained from each patient and genomic DNA was extracted. Coding regions and the intron-exon junctions of the NCSTN gene were amplified by polymerase chain reaction (PCR) according to previously published primer, performing Sanger sequencing on the PCR products. Demographic and clinical data were recorded from every patient.

A total of three unrelated HS/DDD phenotype patients were included in this study. HS (Hurley stage II) and brownish macules in a reticular pattern on the flexural areas were present in all cases. Skin biopsies were performed in each patient and they showed hyperkeratosis with multiple horny follicular plugs and papillary epidermal down-growth with abnormal basal pigment-granule distribution. Case 1 DNA sequencing showed a heterozygous mutation c.1876C>T (p.Arg626Ter) in the NCSTN gene. Case 2 presented a heterozygous mutation c.1325_1326insGTTGTTCTGTAGTGGC (P.Asp443LeufsTer6) of the same gene. These consensus splice site mutations potentially generate aberrant splicing with loss of functionality. These new mutations probably have a pathogenic significance, although there are not included in the mutation database (ClinVar, ESP6500, HGMD). Case 3 DNA sequencing revealed a heterozygous mutation c.1300C>T (p.Arg434Ter) in the NCSTN gene. This variant is described in the ClinVar databases and HGMD (CM123788) as pathogenic associated with HS.

Mutations in NCSTN gene are associated with HS/DDD phenotype. Mutations in other genes such as, PSENEN and POFUT1 have also been reported in HS/DDD phenotype.

064 PS03 - poster session | Expression of chitinase-3-like protein 1 (YKL-40) in lesional and non-lesional skin of hidradenitis suppurativa

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Recently, we suggested chitinase-3-like protein 1 (YKL-40) as a new biomarker for hidradenitis suppurativa (HS) as its serum levels were found to be elevated in patients. The objective of this study was to examine YKL-40 tissue expression in HS.

The study was conducted on 7 patients with HS. The biopsies were obtained from lesional and non-lesional skin. The tissue expression of YKL-40 was assessed using an immunohistochemical method.

Inflammatory infiltrates of various grades were visible in all samples of lesional skin. The inflammatory cells within those infiltrates showed a strong cytoplasmic expression of YKL-40. In non-lesional HS skin the pattern of YKL-40 expression was similar to that observed in healthy skin.

Lesional skin of HS showed enhanced expression of YKL-40. This can argue the role of this molecule in the pathogenesis of the disease and support YKL-40 as valuable biomarker in HS.

065 PS04 - poster session | Novel anti-IL17 antibody (CJM112) reduces inflammation of hidradenitis suppurativa patients in a placebo-controlled trial

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The pathophysiology of hidradenitis suppurativa (HS), a chronic inflammatory skin disease, is not fully understood and the need for effective and safe treatments is still high. This study is a proof of concept study using CJM112 as a novel fully human monoclonal antibody with high affinity to interleukin (IL)-17A.

To test whether the anti-IL17A antibody CJM112 is effective in reducing the inflammatory burden of HS.

Adult HS patients were randomized to receive either CJM112 (300 mg s.c.) or placebo for two sequential treatment periods of 16 weeks in this multicenter, randomized, double blind, placebo controlled trial. Patients were required to have ≥2 affected areas with Hurley stage II and III, ≥4 abscesses and/or nodules, and at least moderate HS-Physician Global Assessment (HS-PGA) to be included. The primary endpoint was the rate of response defined as achieving a 2-point reduction in HS-PGA at week 16.

Patient baseline and disease characteristics were balanced across the two arms (CJM112, n = 33; Placebo, n = 33). Mean serum high sensitivity C-reactive protein (hsCRP) was above normal at baseline (19.2 mg/L and 21.0 mg/L, CJM112 and placebo). Mean β-defensin-2 in serum was higher at baseline (453 pg/mL) than observed in a control Healthy Volunteer group (82 pg/mL).

CJM112 was generally well tolerated and the overall safety profile of CJM112 was similar to placebo group. At 16 weeks, the HS-PGA response rate was 32.3% (10/31) with CJM112, which was significantly superior to 12.5% (4/32) seen with placebo. The decrease in inflammatory lesions (abscesses, nodules and fistulae) was 56% with CJM112 compared to 30% for placebo. A significant difference between treatment groups in mean serum hsCRP was observed at week 16 (10.92 mg/L (CJM112) and 17.18 mg/L (placebo)).

HS has a high inflammatory burden, as demonstrated by increased hsCRP. CJM112 was able to lower significantly this burden as compared to placebo in this 16-week, proof-of-concept trial. To our knowledge, this is the first randomized placebo-controlled trial evaluating the efficacy of an anti-IL-17 antibody in the treatment of HS.

066 PS05 - poster session | Serum levels of psoriasin (S100A7), calgranulin A (S100A8) and calgranulin B (S100A9) in patients suffering from hidradenitis suppurativa

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Hidradenitis suppurativa (HS) is a chronic inflammatory skin disease with high socioeconomic burden. The pathogenesis of HS is still not completely elucidated, but an important role of the aberrant innate immune response has been suggested. Furthermore, there is a lack of serum biomarker for monitoring of patients with HS. Psoriasin (S100A7), calgranulin A (S100A8) and calgranulin B (S100A9) belong to the family of S100 proteins. They show some antimicrobial activity against Escherichia coli, Streptococcus spp. and act as alarmins by priming immune cells for enhanced production of proinflammatory mediators. The aim of the current study was to investigate the serum levels of S100A7, S100A8 and S100A9 in patients with HS compared to controls.

Serum concentrations of S100A7, S100A8, S100A9 were measured in 63 individuals with HS and 31 healthy controls. Commercially available ELISA kit were used. Severity of HS was assessed according

to the Hurley classification. Correlations between serum concentrations of S100 proteins and severity of disease and some other clinical data were evaluated Results: Serum levels of S100A8 and S100A9 were significantly elevated in patients with HS compared to controls ($P = 0.001$ and $P = 0.035$, respectively) and associated with disease severity according to the Hurley staging. Serum level of S100A7 was decreased in patients with HS as compared to controls ($P = 0.025$). S100A7, S100A8, S100A9 may play a role in the pathogenesis of HS. Furthermore, these proteins may constitute the promising biomarker for disease severity monitoring.

067 PS06 - poster session | S100A4 and S100A15 as biomarkers of hidradenitis suppurativa disease activity with potential pathogenetic implications

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Hidradenitis suppurativa (HS) is a chronic, debilitating inflammatory skin disease with the important role of the aberrant innate immune response. There is a lack of suitable serum biomarker that could facilitate monitoring of disease progression and differentiation between stages of disease. S100 proteins constitute a family of small, calcium-binding proteins, that play an important role in the regulation of innate immune mechanisms. Furthermore, S100A4 is extracellular factor with a capacity to influence gene expression by modulation of transcription factors: p53 and NF-κβ. S100A15 is an antimicrobial proteins with proinflammatory activity. The study objective was to investigate serum levels of S100A4 and S100A15 in patients suffering from HS compared to controls.

Serum concentrations of S100A4 and S100A15 were measured in 63 individuals with HS and 31 healthy controls. Severity of HS was assessed according to Hurley classification. Correlations between serum concentrations of S100 proteins and severity of disease and some other clinical data were evaluated. Commercially available ELISA kit were used. Results: Serum level of S100A4 was significantly increased in individuals with HS compared to controls ($P = 0.02$) and associated with disease severity. There was no significant difference in serum levels of S100A15 between group of patients with HS and controls. However, the serum level of S100A15 was significantly elevated in patients with HS Hurley III as compared to controls ($P = 0.0018$).

S100A4 and S100A15 may constitute promising biomarkers for HS, helpful for monitoring of the disease severity. Furthermore, they may play a role in the pathogenesis of HS.

068 PS07 - poster session | The aberrant expression of selected S100 proteins in skin lesions of patients suffering from hidradenitis suppurativa

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Hidradenitis suppurativa (HS) is a chronic inflammatory skin disease. An aberrant expression of certain antimicrobial proteins may play an important role in the pathogenesis of HS. However, this issue is still not elucidated and further investigations are required. S100 constitute a family of small, calcium-binding proteins with some antimicrobial properties that have emerged as key player of innate immunity. Furthermore, expressed by epidermis, they may regulate keratinocyte proliferation. The aim of the study was to investigate expression of S100A7 (psoriasin), S100A8 (calgranulin A) and S100A15 (koebnerisin) in skin lesions and perilesional skin of patients suffering from HS.

Study was conducted on a group of 15 patients suffering from HS. From each individual with HS two biopsies were taken, from lesional and perilesional skin. The expression of S100A7, S100A8, S100A15 (S100A15L, S100A15S) were investigated using quantitative real–time PCR and immunofluorescent analysis. The expression of these proteins in perilesional skin was compared to their expression in healthy skin. The potential associations with diseases activity and some clinical data were assessed. Results: We found overexpression of S100A7 and S100A8 in lesional HS skin as compared to perilesional HS skin and healthy skin ($P < 0.05$).

An aberrant expression of some S100 proteins may play an important role in the pathogenesis of HS.

069 PS08 - poster session | Brazilian consensus of hidradenitis suppurativa

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Hidradenitis suppurativa is a chronic immune mediated disease of universal distribution that causes great damage to the quality of life of the affected individual, whose prevalence is estimated at 0.41% in the Brazilian population. The objective of this work was update on physiopathogenesis, diagnosis and classification of Hidradenitis suppurativa and to establish therapeutic recommendations in the Brazilian reality. It was organized a work group composed of eight dermatologists from several institutions of the country with experience in the treatment of Hidradenitis suppurativa and carried out review on the

topic. Recommendations were elaborated and voted by modified Delphi system and statistical analysis of the results was performed. The Brazilian consensus on the clinical approach of Hidradenitis suppurativa had the support of the Brazilian Society of Dermatology.
A systematic review on clinical treatment was conducted and statements were organized for expert vote. Regarding the use of antibiotics, cyclins and sulfas were chosen as the first line of treatment, and other antibiotics (ciprofloxacin and metronidazole or clindamycin as a second line with caution for rifampicin, whose use should be restricted due to the high incidence of resistant tuberculosis in the country. Agreement greater than 80% among specialists on the use of anti-inflammatory and immunosuppressants as intralesional or systemic corticosteroid and biological as second line was achieved. Recommendations for topical treatment and adjuvant measures, including surgical indication, were established in this consensus.
The document aims to guide the dermatologist to the most appropriate and early treatment of HS in a country where access to therapies is restricted and the epidemiology of diseases is different.

070 PS09 - poster session | Hidradenitis suppurativa: experience in Argentina's reference center

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Hidradenitis Suppurativa (HS) is an immuno-mediated, systemic and chronic inflammatory disease that profoundly affects the quality of life of patients. Objective: To compare the epidemiological characteristics of our population of Hidradenitis Suppurativa patients with the published casuistry. Retrospective work, based on the experience of diagnosis and treatment of 56 patients with HS at the General Hospital of Agudos "Dr. I. Pirovano "during the period 2015-2017. Results: Most frequent affection in women (71.42%). High association with smoking (78.6%), alcohol consumption (89.28%) and obesity (64.28%). 3.5% of the cases had a family history and 46 comorbidities (82%) were observed among all patients, the most frequent being arterial hypertension (26.78%), diabetes mellitus (25%) and dyslipidemias (12.5%). All the patients presented poor quality of life with an average DLQI> 15 and a VAS (pain) that averaged the 8 points. The average age of onset was 24.39 years, with an average delay of diagnosis of 9.75 years. Most of the patients (78.57%) had no diagnosis and in 93% of the cases, it was performed by a dermatologist. 71% of the cases started with lesions in a single anatomical site, the axillary region being the most frequently affected and 29% initially presenting compromise of 2 sites (axillary and inguinal region). We classified 10 patients with Hurley I, 22 patients with Hurley II and 24 patients with Hurley III. Multiple therapies were used without response and only 8/24 severe patients were at the time of consultation with adalimumab.
Although there are many coincidences, items such as age of onset that is later, diagnosis made mostly by dermatologists, most patients attend the consultation in more advanced stages and they are sub-treated.

071 PS10 - poster session | Hidradenitis suppurativa: a single centre experience

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Hidradenitis suppurativa is diagnosed by its clinical features and its chronic nature. Our objective is to describe a clinical characteristic, comorbidities, and response to treatments in patients with hidradenitis suppurativa.
We registered clinical characteristics of patients visited during the period January 2018 to January 2019 at our hospital.
77 patients were visited during this period, 43 were women, and 34 were men. The median age was 38 years. Sixty patients were smokers. Sixteen of these patients were classified as Hurley I, 27 as Hurley II and 28 as Hurley III. 6 patients were not classified with Hurley scale. Eleven patients with severe disease were treated with adalimumab. Most of them achieved a decrease in the inflammatory lesions without adverse events. Eight patients with localized disease were treated with intralesional photodynamic therapy (I-PDT), 6 were man and 2 were women. The tolerance was good, and patients referred a diminution in the inflammatory lesions and a decrease in pain.
We presented our experience of these patients series, their clinical characteristics and their response to different treatments options and how it has affected their quality of life.

072 PS11 - poster session | German consensus on documentation of hidradenitis suppurativa

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Hidradenitis suppurativa (HS) is a chronic inflammatory skin disease which is characterized by the recurrent development of abscesses and fistulae and leads to a severe impairment of the quality of life of the affected persons. In order to improve patient care, optimize the work of care providers, and ensure data comparability, uniform documentation and progress standards are required at the national and international level. This paper compiles the results of the German National Consensus Conference, which dealt with the consensus on standard data sets for HS.
Consensus takes place in several stages, with the data sets being developed through online surveys and subsequent votes/adjustments in the face-to-face meetings. The Federal Consensus Conference was attended by representatives of professional societies and associations, as well as nursing representatives. External experts for HS also took part in the online surveys. The consensus was reached in accordance with the recommendations of the Working Group of the Scientific Medical Societies e.V (AWMF) and the memoranda of

the German Network for Health Services Research (DNVF). After a systematic literature search, the variable items were created and submitted to the experts for approval.

17 representatives of organizations and associations agreed to participate in the consensus building process. A distinction was made between a minimum dataset (MDS) and a standard dataset (SDS). The total of 90 parameters selected by the experts were arranged according to MDS and SDS, with MDS 20 and SDS recording the total of 90 defined parameters. An item was assigned to the MDS if at least ≥ 75% of the experts had found the item necessary. For SDS, the threshold value was at least 50% for necessary and/or desirable. The MDS should be used in every routine examination. The SDS is aimed at specialized centers for HS.

The items were categorized as follows: 1. Basic data, 2. General status and anamnesis, 3. Clinical status of the HS, 4. Diagnostics, 5. Therapy, 6. Patient reported endpoints (PRO), 7. Nutrition and 8. Education.

A standardized documentation of anamnesis, disease progression and therapies enables an improved supply and comparability of the data due to the transparency. The inclusion of patient representatives would be desirable for future consensus conferences. The standard data set should provide the best possible basis for documentation for specialized centers. This does not serve to list all existing measuring instruments and, if necessary, additional parameters useful for certain scientific projects, but represents a documentation set that is as compact as possible in terms of feasibility and efficiency.

073 PS12 - poster session | Hidradenitis suppurativa in the pediatric population: own experience

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Hidradenitis suppurativa (HS) is a chronic and devastating inflammatory disease. Diagnosis of HS is based on clinical features, the disease is more common in women, the first skin lesions usually appear after puberty. The existing literature data concerning children with HS are scarce, therefore the aim of this study was to analyze demographical and clinical data of our pediatric patients suffering from HS.

A retrospective analysis of data collected from pediatric patients treated in our center was conducted. Case series of 10 subjects were analyzed in terms of clinical data, diagnosis and treatment.

The evaluated group consisted of 4 girls and 6 boys aged 13-17 years old (mean 14.5 ± 3.4 years). The severity of disease assessed with Hurley stage I was diagnosed in 2 children, Hurley stage II in 7 subjects and Hurley stage III in 1 patient. Family history of HS in the past, as well as pilonidal cyst were reported in one pediatric patient with HS. In addition, there were no cases of acne conglobate, dissecting cellulitis, inflammatory bowel disease or type 1 diabetes mellitus, both among subjects and their relatives. Four of the subjects (40%) were active smokers and four of the subjects' mothers (40%) smoked cigarettes during pregnancy. The mean Body Mass Index (BMI) of the patients was in the 96 percentile what represents obesity. 90% of skin changes in children were located both in the axilla and groin area. All of the patients received systemic antibiotic treatment, half of them have not achieved marked clinical improvement.

HS is still a major therapeutic challenge, due to poor response to the recommended oral treatment. Exposure to tobacco and obesity are possible HS risk factors in this group of patients as majority of them were overweighted and some of them were smokers. Although none of the patients developed metabolic syndrome, it might appear further in time due to its strong association with HS among adults. The role of education of young patients is crucial, since reduction of body weight and smoking cessation may support oral pharmacological treatment.

074 PS13 - poster session | Which factors determine affected sites in hidradenitis suppurativa?

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Clinical presentation of hidradenitis suppurativa (HS) is protean regarding severity and affected sites. Factors influencing the site of occurrence are still misunderstood.

To evaluate the clinical factors that influence which sites are affected in HS in a large cohort of patients.

The chart of patients with HS and complete data about demographics (gender, age, IMC, family history), disease severity (Hurley stage), smoking and comorbidities (joint and bowel disease (IBD), acne vulgaris, acne conglobata, pilonidal sinus disease, dissecting folliculitis of the scalp) were included. Logistic regression analyses were performed to identify factors able to negatively (the risk of site involvement is higher if the factor is absent) or positively (increased risk when the factor is present) predict whether a site is affected independently of other factors.

We included 1138 patients with complete data (female: 64%; mean age: 32 years; family history: 25%; smoking: 79%; Hurley I/II/III: 56%/31%/13%; mean age at disease onset: 21 years). Female gender was an independent predictor of breast, genital, and groin involvement. Male gender was an independent predictor of neck, perineum and intergluteal involvement. Age at onset was an independent negative predictor of genital involvement. Age at the study was an independent predictor of gluteal, groin, perineum, and supra-pubic involvement. BMI positively correlated with breast, armpit, inner tight, supra-pubic

and intergluteal involvement, and negatively with gluteal, groin, anal, and retro-auricular involvement. A family history was an independent predictor of neck and gluteal involvement. Smoking was positively associated with genital and retro-auricular involvement. Hurley stage was an independent predictor of armpit, perineum, and intergluteal involvement. Although joint disease was not associated with any of the sites, the association of IBD with HS positively correlated with anal involvement, and negatively with neck and retro-auricular involvement. A strong association was observed between acne vulgaris, dissecting folliculitis of the scalp and involvement of the neck and the retro-auricular region. This result is consistent with the follicular occlusion triad but not with the tetrad suggesting different mechanisms between pilonidal sinus disease and the triad. Some sites were associated with areas in the vicinity (ex: genital and groin involvements; concurrent involvement of buttocks, sub-gluteal folds and inner tights). Both genital and anal involvements were independently associated with perineum involvement but they did not correlate with each other suggesting that the generic 'anogenital involvement' may not be relevant.

Associations can clearly be inferred between clinical factors and affected sites. Further analyses are mandatory to use these associations to define patient subgroups and to reflect upon pathophysiology of the disease.

075 PS14 - poster session | A clinico-epidemiological study of hidradenitis suppurativa in Greece: data from three university centers of reference (2016-2018)

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Hidradenitis suppurativa (HS) is a chronic, recurrent, debilitating, disease of apocrine gland-bearing areas of the skin. It affects 1-2% of the population causing significant morbidity and impairing quality of life. We sought to depict the epidemiological and clinical characteristics, as well as risk factors and comorbidities among HS patients in Greece.

Adult patients diagnosed with HS, attending the HS Clinics of three academic dermatology departments in Athens and in Thessaloniki during a three-year period (2016-2018). For all patients, demographic characteristics, risk factors, history, clinical parameters, quality of life assessment and previous and current treatments, were recorded in the context of a national HS registry. The software SPSS, version 22.0 was used for the statistical analysis of the data.

In total, 208 patients were studied. Of them, 53.8% (n = 112) were females. The mean age was 36.67 ± 12.84 years, the mean age at HS onset was 25.48 ± 11.40, and the mean age at HS diagnosis was 31.80 ± 11.87 years. Forty six patients (22.1%) had a family history of HS, 74.5% were current smokers, 69.2% had an abnormally increased BMI, and 27.9% reported a history of a pilonidal cyst. In the majority of the patients (n = 125), the disease remained localized in its initial presentation site, the groin and the axilla being the most frequently affected areas. Hundred-sixty patients (76.9%) had more than one affected areas; the groins, the axillae and the buttocks were the most commonly affected regions. Regarding Hurley staging, 27.9% of the patients were classified as Hurley stage I, 30.3% as Hurley stage II, and 41.8% as Hurley stage III. Comorbidities were present in 64.9% (n = 135) of our patients. The most common comorbidity was acne (46.9%), followed by endocrinal/metabolic disorders and depression. The mean DLQI score was 12.33 ± 8.58. Almost half of the patients (n = 90) had undergone a surgical procedure at any time during the disease course. At the present time, the majority of our patients (39.9%) are under biologic therapy (adalimumab), followed by topical treatment (28.4%), and oral antibiotics (25.9%).

In our cohort, there was only a slight female preponderance (53.8%). Current smoking and obesity were very prevalent. A significant delay in HS diagnosis (mean 6.32 years) was documented. Family history of HS was common. The groins and the axillae were the most commonly affected sites, and the majority of patients had more than one affected areas. Comorbidities were present in two-thirds of the patients and over one-fourth had history of pilonidal sinus. Most patients (72.1%) were classified as Hurley stage II or III. A moderate to severe impairment of quality of life was observed (DLQI). Surgical interventions were common during the course of the disease. Currently, 65.8% of our patients are under systemic medical therapy, mostly biologic therapy with adalimumab.

076 PS15 - poster session | Hidradenitis suppurativa in a cohort of children and adolescents with overweight and obesity

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Hidradenitis suppurativa (HS) is a chronic, inflammatory, and recurring disease, mainly observed in adults. Obesity is considered an important independent factor in HS development and associated with a higher prevalence of HS in children as well. The objective of this study was to characterize the clinical presentation of HS in children/adolescents with overweight/obesity.

We performed a cross-sectional observational study during January 2007 – April 2015. Patients with overweight/obesity (5-17 years

of age, BMI > 90th percentile) referred to The Children's Obesity Clinic, Department of Paediatrics, Copenhagen University Hospital Holbæk, Denmark, underwent screening for dermatological conditions. A dermatologist ascertained the diagnosis of HS, and disease severity was assessed using Hurley staging, and Sartorius score. Tobacco smoke exposure, body mass index (BMI) standard deviation score (SDS), and psychiatric comorbidities were recorded.

A total of 195 children/adolescents underwent screening for dermatological conditions. Nine patients screened positive, and six of these patients were available for examination. Five of six patients presented with HS, corresponding to a dermatologist-verified prevalence of 2.6 % (confidence interval-95%: 0.1-5.9%). All HS cases were mild (median Sartorius score of 9). Four of five patients (with varying constellations) reported tobacco exposure, a positive family history of HS, and exhibited psychiatric comorbidities.

Our findings suggest a high prevalence of HS in children/adolescents with overweight/obesity, and support associations with tobacco exposure and a familial disposition to HS. We found a strong association with psychiatric comorbidities, suggesting that children/adolescents with overweight/obesity would benefit from routine screening for HS and psychiatric comorbidities.

077 PS16 - poster session | Demographic and biometric data of 1428 patients with hidradenitis suppurativa: EpiVer French multicenter study

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Hidradenitis suppurativa(HS) is considered an uncommon and almost rare disease. French series are few.

As part of a French network that studies HS (RésoVerneuil) we wanted to identify the demographic and biometric characteristics of patients with HS in a multicentric study not biased by predominant or exclusive hospital recruitment.

Since March 2016 to December 2017 RésoVerneuil members have anonymously registered the standardized examination data of their patients with HS as part of the EpiVer study. 1428 subjects have been identified: 884 women (61.9%) and 544 men (38.1%).

Most patients were young female. The most frequent age range was between 20 and 39 years (63.72%). HS was rare before the age of 15 (0.91%) and after the age of 59 (2.1%). Mean weight was 79.43 kg (median 76, standard deviation 19.67). The mean body mass index (BMI) was 27.58 (median 26.2, standard deviation 6.47). Mean abdominal perimeter was 95.36 (median 93, standard deviation 17.41). 29.20% of the subjects were overweight (BMI≥25 and <30) and 28.62% were obese (BMI≥30), of whom 4.41% had pathological obesity (BMI≥40) and 13.28% were potentially eligible to bariatric surgery (BMI> 35 with comorbidity). Since the Obépi study on the entire French population found overweight in 32.3% of French and obesity in 15% of French, our study confirmed that patients suffering from HS are fatter than the average French. A relationship between overweight and HS has not been demonstrated, but overweight and pathological obesity are frequently associated with HS. A multidisciplinary collaboration with overweight medical and surgical specialists and with cardiologists is essential for the overall care of these patients.

078 PS17 - poster session | Personal and family history of 1428 subjects with hidradenitis suppurativa: EpiVer study

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Epiver survey is a French multicenter study and one of the largest series of patients with Hidradenitis Suppurativa (HS)non-retrieved from health insurance databases and diagnosed by experts.

As part of a French network that studies HS (RésoVerneuil), we wanted to characterize the personal and family history of our patients with HS. These data came from the Epiver survey.

From March 2016 to December 2017 RésoVerneuil members anonymously recorded data of their patients with HS. Personal history (pilonidal sinus (PNS), acne, acne treated with isotretinoin), cardiovascular history (arterial hypertension, stroke/transient ischemic attack (TIA), angina/myocardial infarction, dyslipidemia, insulin-dependent diabetes, non-insulin-dependent diabetes), inflammatory disease history (inflammatory bowel disease (IBD), inflammatory rheumatism (IR)) and familiar history (HS, PNS, IBD, IR) were investigated.

The incidence of familiar and personal history of IBD and IR was particularly high. It is considered that 0.5% of the French population is suffering from IBD against 3.5% of our patients. 10% of people with IBD have a first degree relative with IBD, whereas 4.7% of our patients and IBD-free population have a direct relative with IBD. The incidence of personal history of IR was 5.1%, whereas it is assumed that the incidence of ankylosing spondylitis would be 1% in the general population. Regarding risk factors for cardiovascular disease (hypertension, dyslipidemia, diabetes) there was an unusually.

079 PS18 - poster session | Evaluation of medical and surgical treatments for hidradenitis suppurativa from the Scandinavian registry (HISREG)

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Hidradenitis suppurativa (HS) substantially affects health-related quality-of-life outcomes. Most treatment options are supported by low quality of evidence without validated outcomes. The aim of this study was to evaluate the efficacy of surgical and medical interventions using physician- and patient-reported outcomes registered in the prospective Nordic Registry for HS (HISREG).

Data were extracted for all adult patients registered in HISREG between January 2013 and April 2016. Primary endpoints included Dermatology Life Quality Index (DLQI) scores, pain as measured using a numeric rating scale (NRS), Sartorius score, and Hurley classification. Minimum clinically important differences (MCIDs) for DLQI and NRS pain were analyzed. Secondary endpoints included comparisons among different treatment groups, safety, and complications of various treatments.

Two hundred fifty-five patients were included in the study: 31, 188, and 36 patients had Hurley stages I, II, and III disease, respectively. Treatment with CO2 lasers was the most common treatment modality. One hundred forty-nine patients (58.4%) were treated with surgical intervention, 87 (34.1%) received antibiotics and/or anti-inflammatory treatments, and 19 (7.5%) were treated with both surgery and medical intervention. No patients received biologic treatment. In patients with surgical treatments, Sartorius scores were significantly improved compared with baseline ($P = 0.001$), 83 patients (55%) achieved a DLQI MCID, and 75 patients (49.7%) achieved an NRS pain MCID. In patients with medical treatments, Sartorius scores were not significantly improved compared with baseline ($P = 0.582$)

25 patients (28%) achieved a DLQI MCID and 28 patients (31%) achieved an NRS pain MCID. In patients treated with surgical and medical combination, 9 (48%) achieved DLQI and NRS pain MCIDs and Sartorius scores were significantly improved.

CO2 laser treatment is more effective than the non-biologic medical treatments in this analysis based on physician- and patient-derived outcomes. The study provides limited evidence for the combination of medical and surgical therapies in patients with HS.

080 PS19 - poster session | Hidradenitis suppurativa: still unknown in primary care assistance

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Hidradenitis suppurativa (HS) is a chronic recurrent inflammatory skin disorder that involves hair follicles with apocrine glands, typically localized at intertriginous areas. HS is uncommon in children, with onset generally occurring after puberty. Prepubertal onset-before 11 years of age-is estimated to occur in 2% of patients with HS (7,7% under 13 years old), being the disease still unknown and underdiagnosed by primary care professionals. Children with HS are more likely to have a hormonal imbalance, a positive family history and a wider-spread affectation than adults with HS. Regardless of age of presentation, HS impacts quality of life, functioning and self-esteem.

The aim of this study is to evaluate the knowledge about Hidradenitis suppurativa in the primary care group.

A survey was performed to primary care physicians including 5 questions

1. Hidradenitis suppurativa is: an infectious condition or an inflammatory condition.
2. Hidradenitis suppurativa diagnosis in children under 12 years old is, in my opinion: easy or difficult.
3. The most frequent age of involvement of Hidradenitis suppurativa in children is: under 4 years old, between 5 and 11 years old or between 12 and 17 years old.
4. How many patients with Hidradenitis suppurativa have you visited in the last year?
5. What Medical Specialist do you refer to patients with Hidradenitis suppurativa?

A total of 36 primary care physicians answered the survey. Regarding the first question; half of the respondents answered that HS is an inflammatory condition, 25% that it is an infectious process and 25% didn't answer the question. To the question number 2; 42% answered that the diagnosis of HS in children under 12 years old is difficult while 30% answered that it is easy, 4 of which had answered in the first question that HS is an infectious process. 28% didn't answer the question. To the third question; 67% said that most frequent age of involvement of HS is between 12 and 17 years old, 8% between 5 and 11 years old and 25% didn't answer. To the question number 4; 30% of the respondents had not visit any patient with HS in the last year, 14% had visited only one patient, 11% two patients, 3% 3 patients, 3% from 5 to 6 patients, 3% more than 10 patients and 36% didn't answer the question. To question number 5; only 17% refer patients with HS to Dermatology Specialists. 14% refer them to General Surgery, 9% to Dermatology or General Surgery indistinctly and 3% to Maxillofacial Surgery. 11% of the respondents don't refer patients to any Medical Specialist and treat them by themselves. 46% didn't answer the question.

Little is known about HS in the primary care group. Proper training on HS in primary care physicians is needed in order to do an early

diagnosis given the possibility of stopping disease progression if an appropriate treatment is established during the early stages of the disease.

081 PS20 - poster session | Is subtyping of mammary HS lesions possible: a case series

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Different clinical sub- and phenotypes of HS has been presented in the literature. Van der Zee and Jemec suggest six subtypes (the regular type, frictional furuncle type, scarring folliculitis type, conglobata type, syndromic type, ectopic type) whereas Canoui-Poitrine et al. identified three different subgroups of HS (Gluteal, Follicular, Axillary-mammary). External mechanical stress has also been described as a factor contributing to HS, a clinical subgroup with 'Koebnerization'. The purpose of subtyping is more than describing a disease as it potentially contributes to a more tailored personalized treatment. Based on our own clinical observations, we have noticed that HS patient with involvement of the breast area tend to show four separate morphologies. The first is characterized by deep-seated nodules and/or boils in the lower quadrants of the breasts, which may later perforate the skin causing suppuration. The second, by ropelike scarring at the intermammary area above the sternum (pilonidal cyst–like). The third by nodules at places exposed to pressure from bra straps and areas of friction. The fourth is a mixture of the tree.

To validate our observation we therefore recorded the morphology and anamnestic data of patients with HS involvement of the mammary area. All patients were recruited at Dept. of dermatology at Zealand University Hospital, Roskilde during consultations in our outpatient clinic.

We included 20 patients in this short case series overview. Half of the 20 patients (10/20) showed characteristics for one group, the spontaneous type (N = 6) being the most prevalent, friction (N = 3) and sternum type (N = 1). A single patient did not correspond to any group as she only had folliculitis. In the mixed group (n = 9), two patients actually had all three morphologies.

The clinical importance of sub-classifying HS based on morphology should be significant if the different phenotypes either arise due to different etiologies or could possibly have different prognosis or response to treatment. In our study, we present four different morphologies of HS targeting the breast and show that most of the patients have mixed types. Interestingly there seemed to be a high co-occurrence of the spontaneous and the frictional type, as more than 63 % (7/11) of the patients showing the friction morphology also had the spontaneous morphology. It may be that for most, the friction type is a secondary development based on Koebnerization in patients with the more common spontaneous type. One may then imagine that the three patients showing only the friction type were more susceptible to Koebnerization based on higher BMI. However this was not the case as they had an average BMI of 33.3 whereas the average BMI of the other 17 patients were 34.8 (*P*-value 0.69). More data is needed to better elucidate if there is a difference between our four groups, and to justify if such a grouping is sound.

082 PS21 - poster session | Medical and surgical treatment of obesity

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The prevalence of obesity has reached epidemic proportions. Conceptualization of obesity as a chronic disease facilitates greater understanding its treatment. The NIH Consensus Conference on Gastrointestinal Surgery for Severe Obesity provides a framework by which to manage the severely obese--specifically providing medical versus surgical recommendations which are based on scientific and outcomes data. Medical treatments of obesity include primary prevention, dietary intervention, increased physical activity, behavior modification, and pharmacotherapy. Surgical treatment for obesity is based on the extensive neural-hormonal effects of weight loss surgery on metabolism, and as such is better termed Metabolic Surgery. Surgery is not limited to the procedure itself, it also necessitates thorough preoperative evaluation, risk assessment, and counseling. The most common metabolic surgical procedures include Roux-en-Y gastric bypass, adjustable gastric band, sleeve gastrectomy, and biliopancreatic diversion. Surgical outcomes for metabolic surgery are well studied and demonstrate superior long-term weight loss compared to medical management in cases of severe obesity. The prevalence of obesity has reached epidemic proportions. Conceptualization of obesity as a chronic disease facilitates greater understanding its treatment. The NIH Consensus Conference on Gastrointestinal Surgery for Severe Obesity provides a framework by which to manage the severely obese--specifically providing medical versus surgical recommendations which are based on scientific and outcomes data. Medical treatments of obesity include primary prevention, dietary intervention, increased physical activity, behavior modification, and pharmacotherapy. Surgical treatment for obesity is based on the extensive neural-hormonal effects of weight loss surgery on metabolism, and as such is better termed Metabolic Surgery. Surgery is not limited to the procedure itself, it also necessitates thorough preoperative evaluation, risk assessment, and counseling. The most common metabolic surgical procedures include Roux-en-Y gastric bypass, adjustable gastric band, sleeve gastrectomy, and biliopancreatic diversion. Surgical outcomes for metabolic surgery are well studied and demonstrate superior long-term weight loss compared to medical management in cases of severe obesity.

083 PS22 - poster session | Frictional hidradenitis suppurativa induced by prostheses: an exogenous trigger?

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Although the pathophysiology of Hidradenitis Suppurativa (HS) is not fully understood, there have been many well described contributing factors; family history, obesity and smoking. Mechanical stress on the skin is linked to HS. There have, however, been few documented case reports illustrating the direct impact of external frictional sources as a cause. We present two cases in which the development of HS is directly linked to friction by exogenous triggers.

A 46 year old gentleman presented with recurrent abscesses in his left groin. He had previously undergone a left above-knee amputation following trauma and was using a lower limb prosthesis. He reported the abscesses had not been present prior to the amputation. He was of normal BMI and had no family history of HS, but did have a 20 pack year smoking history. Abscesses were isolated to the left groin area, the area in direct contact with his prosthesis. He had previously undergone a surgical washout of this area but had not undergone any medical treatment prior to this presentation.

The second patient, a 49 year old gentleman, presented with recurrent bilateral, axillary abscesses. He had a background history of arthrogryposis multiplex congenita, a congenital disorder characterised by multiple contractures of the joints requiring the use of axillary crutches to mobilise. He had no family history of HS, was of normal BMI and was an ex-smoker. He had previously undergone treatment with doxycycline and lymecycline. He had also undergone surgical washouts of abscesses. His symptoms were isolated to his axillae, areas directly in contact with his crutches. He was diagnosed clinically with HS.

The development of HS in both of these patients was isolated to the areas directly susceptible to friction from the external agent; in case one, the lower limb prosthesis and in case two the use of axillary crutches. A recent case series by J. Boer described the development of HS at ectopic sites exposed to friction. He queried whether HS should be included in those dermatoses that exhibit the Koebner Phenomenon. It is likely that in susceptible patients, the mechanical friction and also the warm microclimate associated with prostheses promotes the development of HS.

084 PS23 - poster session | A case with fatal outcome due to squamous cell carcinoma over perineal hidradenitis suppurativa

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A 57-year-old man was first diagnosed with hidradenitis suppurativa predominantly on buttocks when aged 24 years progressing to Hurley III. Since then he was treated with topical and oral antibiotics, intralesional corticosteroids, surgery in many occasions (wide resections), anti TNF (infliximab and adalimumab) and ustekinumab with poor control of the disease. The histopathological study of the last resection sample (at age 54) showed an infiltration of skin and lymph nodes by an squamous cell carcinoma with HPV genotyping negative. He received palliative chemotherapy (neither wide surgical excision nor radiotherapy were feasible) but finally died

Hidradenitis suppurativa (HS) is a chronic inflammatory condition affecting the apocrine glands of the axilla, groin, and perianal region. Although it is a common condition, it is rarely associated with squamous cell carcinoma (SCC). The prevalence of SCC associated with HS is approximately 4.6%, and is more common among men. It is likely that the chronic irritation and inflammation characteristic of HS drive the malignant transformation to SCC, and recent work has identified other potential risk factors for this malignant transformation, including human papillomavirus (HPV) infection and tobacco use. There have been around 80 reports of this uncommon complication of HS in the literature and approximately half of the patients succumbed to their disease, being the grade of carcinoma the only predictor of mortality. The management of SCC in HS has not been standardized, but current reports recommend aggressive surgical excision with at least 2 cm margins.

This new case underlines the importance of close follow-up and aggressive management of patients with HS because the consequences can be devastating.

085 PS24 - poster session | Association of hidradenitis suppurativa and keloid formation: a therapeutic challenge

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Keloids are benign fibroproliferative tumors that extend beyond the margin of a cutaneous trauma site. The association between lesional HS and keloids have never been described before. Our objective is to study the characteristic of those patients who developed keloids on top of the HS lesions and discuss the therapeutic challenges associated with this condition.

We report nine cases of keloids arising from Hidradenitis Suppurativa lesions. All males and females HS patients with HS keloidal lesions above the age of 12 were included. Patients with keloids in other than the classic HS sites e.g. ear were excluded.

We identified 8 females and 2 males. In our group, the 9 patient ethnicities included African (3), Asians (2), Middle Eastern (2) and Caucasians (1). The patients' ages ranged from 25-63 years. The most frequent site was the chest (in 60% of the cases), with one patient having extensive keloids in multiple sites. The severity of the HS using Hurley staging methods included mild to severe disease. In our study, 4 patients had stage I, 2 patients had stage II, and 3 patients had stage III disease. Our 9 patients were either treated with

intralesional injection of triamcinolone or had received no specific treatment for the keloids. All patients received treatment for HS that included 3 out of 9 patients received Adalimumab for the treatment of HS.

Keloid formation in patients with hidradenitis suppurativa (HS) likely results from local tissue inflammation secondary to HS inflammation. However, this poses a significant challenge both for surgeons and dermatologists. The management of keloids in HS patients can be difficult given the chronicity of the skin inflammation and the formation of deep tunnels and tracts. The majority of our cases were treated by intralesional triamcinolone. However, a good response was observed to immunosuppressive therapy i.e. adalimumab in two reported cases, and this approach deserves further investigation. We recommend a trial of intralesional triamcinolone concomitantly with immunosuppressive e.g. adalimumab and antibacterial therapy. Surgery should be used as a second line approach, followed by silver sheeting while patients are on HS immunosuppressive therapy. The addition of radiation should be reserved for refractory keloids. Further studies are necessary to understand the pathogenesis of the keloids in the setting of HS.

086 PS25 - poster session | Association between hidradenitis suppurativa and myotonic dystrophy: a case report

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Myotonic dystrophy (Steinert's disease) is the commonest form of muscular dystrophy in adults. It is inherited as an autosomal dominant trait and caused by an unstable expansion of a trinucleotide. Circulating androgens are decreased in myotonic dystrophy compared with normal controls. The association between an androgen-dependent disease, as is hidradenitis suppurativa (HS), and myotonic dystrophy is relatively frequent.

We present the case of a 44 year-old woman with known androgen-dependent condition (HS), in the setting of a disease associated with low circulating androgens (Steinert´s disease). She had family history of myotonic dystrophy (grandmother, mother and a sister) and was diagnosed of idiopathic hirsutism and Hurley III HS in her puberty. She presented multiple inflammatory nodules, abscesses and fistulae in axillae, groins and especially in submammary folds. She had multiple surgeries in axillae and groins and for the last eighteen months has been treated with adalimumab 40 mg/week with good response and almost total control of the disease.

Available literature provides important clinical evidence for abnormalities of end-organ sensitivity to androgens in myotonic dystrophy and also provides insight into the mechanism of androgen-dependent diseases.

Some authors support the hypothesis that the peripheral response to androgens rather than absolute circulating levels of androgens is important in androgen-dependent conditions as HS.

087 PS26 - poster session | Is psoriasis a comorbidity of hidradenitis suppurativa?

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In traditional dermatology we have focused on the fact that psoriasis (PSO) is a disease of the epidermis, and hidradenitis suppurativa (HS) is a disease of the hair follicles. The connection between the two diseases is known in rare syndromes such as PsAPASH and SAPHO. Results from immunological investigations of HS skin has demonstrated an upregulation of IL-12/23 and TNFα which as a common pathogenic link between the diseases. Furthermore, the idea of a common mechanisms is lent credence by the fact that both PSO and HS patients show effect of treatment with IL-12/23 inhibitor ustekinumab and TNF α inhibitors (infliximab, adalimumab).

To explore a possible relationship between the diseases we recorded the number of all patients attending the outpatient clinic at the Dept. of Dermatology, Zealand University Hospital, Roskilde, Denmark for the diagnosis HS (ICD10 DL73.2) or PSO (ICD10 DL40.0, DL40.3, DL40.4, DL40.8 and DL40.9). The numbers that had HS as well as one of the PSO diagnoses were compared with recent data from the Danish blood donor study (DBDS) examining the prevalence of HS cases and a study examining the psoriasis prevalence in Denmark.

A total of 620 PSO patients and 440 HS patients were included of which 24 of 1,036 patients had both diagnoses. This means that 5.45 % of all our HS patients also had PSO, and 3.87 % of our PSO patients had HS. We then performed Chi-square test compared this to the 1.8 % of Danish blood donors having HS and 2.2 % of Danes having psoriasis. In both instances, the differences were statistically significant ($P = 0.00015$ and $P < 0.0001$).

We found a strong indication that HS and PSO may be associated as patients with HS had PSO 2.46 times as often as would be expected and patients with PSO had HS 2.15 times as would be expected when compared with the background population.

Such a connection could possibly be a common pathway governed by the increased secretion of IL-12/23 and TNFα cytokines.

088 PS27 - poster session | Is severity of hidradenitis suppurativa related to hypertension and angina pectoris? EpiVer study on 1428 subjects

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Epiver is a French multicenter study and one of the largest series of patients with Hidradenitis Suppurativa (HS).

To evaluate the relationship between the presence of hypertension and angina pectoris and the severity of HS with regard to the impact on quality of life, pain and stage of the disease.

From March 2016 to December 2017 ResoVerneuil members assessed quality of life (DLQI scale), pain (Visual Analogue Scale, VAS) and presence of hypertension and angina pectorisin patients with HS. Results: 11 patients had angina pectoris and 99 had hypertension. In presence or absence of hypertension, average DLQI was 13 and 12 respectively, and average VAS was 5. In presence or absence of angina, average DLQI was 15 and 12 respectively and average EVA was 5. In presence or absence of hypertension, Hurley stage I was 26.5% and 45.3%, stage II was 46% and 39.7 % and stage III was 27.5 % and 15% respectively. In presence or absence of angina pectoris, Hurley stage I was 36% and 44%, stage II was 18% and 40 % and stage III was 46 % and 16% respectively. Conclusions: Quality of life and pain were not much influenced by the presence of hypertension and angina pectoris. However, Hurley stage increased in case of hypertension and angina pectoris. The interpretation of these data must be careful considering the small size of the population with angina pectoris and hypertension. Although our subjects with hypertension and angina pectoris were few, it should be considered that they were found in a population with a median age of 31 years, and this aspect should raise questions about a relationship between HS and cardiovascular disease that could be explained by the inflammatory reflex.

089 PS28 - poster session | Ustekinumab with intravenous induction: results in hidradenitis suppurativa

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Adalimumab is the only approved biological therapy for hidradenitis suppurativa (HS), but primary and secondary failures have been reported. Last published recommendations support the use of infliximab, anakinra and ustekinumab as alternative off-label biologic therapies in HS. Ustekinumab has a 4C evidence level, and has commonly been used in the dosages recommended for Crohn´s disease.

We performed a multicentric retrospective review of patients with HS treated with the intravenous infusion adjusted by weight (260 to 520 mg) followed by subcutaneous maintenance dose of 90 mg every 8 weeks, as recently approved for Crohn's disease. The minimal follow-up period required for inclusion was 16 weeks.

14 patients from six Spanish hospitals were included. Six of them had a concomitant Crohn's disease, and in 8 of them ustekinumab was approved as off-label treatment. In 50% of them, therapeutic objectives, measured by means of HiSCR, DLQI and VAS of pain, were reached at week 16. In 71.42% of treated patients DLQI and VAS of pain improved, irrespective of achievement of HiSCR. Only 2 patients abandoned treatment due to lack of efficacy or patient's preferences. Combination treatment, being the most frequent corticosteroids and antibiotics, was needed in order to treat intercurrent flares in 5 patients. No adverse effects related to ustekinumab were reported.

With the limitations inherent to a retrospective review, and a limited follow-up period, the therapeutical regime of ustekinumab with an initial weight-adjusted intravenous induction followed by subcutaneous administration of 90 mg every 8 weeks was safe and moderately efficient in treating HS with failure to previous biologic therapy. Ideally, efficacy of ustekinumab in HS should be tested in randomized and controlled clinical trials.

090 PS29 - poster session | Interruption of oral clindamycin plus rifampicin therapy in patients with hidradenitis suppurativa: an observational study to assess prevalence and causes

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To assess the prevalence of clindamycin-rifampicin treatment interruption in patients with hidradenitis suppurativa (HS).

Retrospective, observational, descriptive, multicenter study. Inclusion criteria were adults (≥ 18 years) with HS, Hurley stage II or III, who received clindamycin plus rifampicin for the first time. We excluded patients receiving other systemic therapy for HS simultaneously.

We included 509 patients from 14 Spanish hospitals. 272 (53.4%) were female, 344 (67.6%) were obese/overweight (BMI≥25 kg/m^2) and 378 (74.2%) were classified as Hurley II. A total of 135 (26.5%) patients interrupted their treatment with clindamycin plus rifampicin, 100 of them (74.1%) due to adverse events. Variables correlated to higher treatment interruption prevalence were age of 50 years or older (adjusted odds ratio [ORa] 1.8, 95% confidence interval [CI] 1.0–3.1, $P = 0.048$) and smoking (ORa 1.7, 95% CI 1.0–2.7 $P = 0.033$). There were 182 adverse effects in 145 (28.5%) patients, including 108 (21.2%) gastrointestinal (GI) effects and 19 (3.7%) instances of non-specific pain. None of the

patients with GI disturbances presented Clostridium difficile colitis, and all responded well to conservative management.

Treatment interruption and adverse event prevalence were higher than previously described. The main reason for interrupting treatment was adverse events. Older age and smoking were both associated with the outcome. Physicians may consider prescribing probiotics in HS patients at higher risk for GI side effects.

091 PS30 - poster session | Effectivity and safety of adalimumab in hidradenitis suppurativa

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Adalimumab has emerged as an effective therapy in patients with hidradenitis suppurativa unresponsive to other medical therapies.

Objectives of our study is to assess effectivity of adalimumab in a "real-world" setting. A single-center retrospective study was undergone in Dermatology Department of General University Hospital in Valencia. Patients with hidradenitis suppurativa treated with adalimumab between January and December 2017 were selected. Baseline clinical records and HiSCORE at week 12 were retrieved.

19 patients were studied. 11 were male and 8 female. Mean age was 35,2 years. All patients continued treatment till week 12 and no loses were observed. HiSCR was achieved by 13 patients and unachieved by 6 patients. Treatment was administered alone in 7 patients, concomitant with surgery in 5 patients (1 month before or after surgery) and before surgery in 7 patients (two or more months before surgery). Any patient presented any significant adverse event during treatment.

Adalimumab is safe and effective in patients with hidradenitis suppurativa. It can be administered as stand-alone therapy or in combination with surgery of chronic lesions.

092 PS31 - poster session | Comparisons of disease status and symptom remission of hidradenitis suppurativa patients treated with biologics and non-biologics

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To examine the prevalence of symptoms in various locations and compare the disease remission among hidradenitis suppurativa (HS) patients treated with biologics and non-biologics from disease diagnoses to most recent consultation.

A multi-centre medical chart review study of HS patients was conducted among dermatologists in clinical practices in France, Germany, Italy, Spain and UK (5EU) to collect de-identified data on their prescribing history in 2Q 2018 (2Q18).. Physicians were screened for practice-duration (<30 years), patient-volume (see >4 moderate/severe HS patients/quarter) and history of biologic prescription for any condition. Physicians were recruited from a large panel to be geographically representative in each country. Patient charts of 4 + successive HS patients visiting each centre/practice were selected. Data on demographics, disease characteristics (including symptom improvement/disease remission), and treatment history were captured. Appropriate statistical analyses for comparison between the two groups were conducted.

In 2Q18, 226 physicians abstracted a total of 904 HS patients (448 on biologic treatment, 456 on non-biologic treatment). At diagnosis, 78.8% of biologic patients and 79.8% of non-biologic patients presented with HS symptoms in the armpit area; decreasing at the most recent consultation to 57.6% ($P \leq 0.01$) and 67.8%, ($P \leq 0.01$) respectively. 73.9% of biologic patients and 73.0% of non-biologic patients presented in the groin/genital area at diagnosis, with both groups decreasing (58.3%, 63.8% respectively; both $P \leq 0.01$). Decreases were also observed in the buttock area (biologic patient at diagnosis: 55.8%; at consultation: 33.7%, $P \leq 0.01$; non-biologic patient at diagnosis: 41.2%; at consultation: 30.0%, $P \leq 0.01$), as well as the anal/perianal area (biologic patient at diagnosis: 52.9%; at consultation: 31.5%, $P \leq 0.01$; non-biologic patient at diagnosis: 35.8%; at consultation: 26.3%, $P \leq 0.01$). The biologic group also displayed decreases in all other body locations since diagnosis (breast(s), 27.7% vs 13.0%; back, 18.1% vs 10.5%; legs, 15.6% vs 9.4%; stomach, 8.0% vs 3.6%; all $P \leq 0.01$), however, similar reduction was not seen in the non-biologic group. The non-biologic group showed a decrease of symptoms in the breast(s) area (22.6% vs 17.3%; $P \leq 0.05$), but all other areas did not show a significant decrease over this time period (back, 10.8% vs 7.5%; legs, 5.9% vs 5.9%; stomach, 2.9% vs 2.4%).

In this study, both biologic and non-biologic patient groups showed overall improvement in HS symptom presentation from diagnosis to their most recent consultation. However, the biologic group showed a significant reduction in HS symptoms across all assessment areas. This demonstrates the increased efficacy of biologic treatment of HS in comparison to non-biologic treatment, facilitating the faster improvement in patient quality of life.

093 PS32 - poster session | Can hidradenitis suppurativa targeted antibiotic strategy improve Crohn disease symptoms?

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Crohn Disease (CD) can be associated to Hidradenitis Suppurativa (HS) in approximately 2% of cases. Pathophysiology of these diseases has similarities, with involvement of inflammatory dysregulation, genetics and microbiome. Use of antibiotics in CD is limited, but common, notably in perianal CD. Targeted antibiotics are a possible

treatment in HS. We studied the impact of targeted antibiotics prescribed for HS on CD symptoms in a small series of patients with associated CD and HS.

We retrospectively studied the evolution of CD symptoms and in particular the number of diarrheal stools per day in 11 consecutive CD+HS patients before and after a targeted antibiotherapy for HS.

11 patients were included, 9 females and 2 males, 9/11 were smokers. Mean age was 40 years old. Median disease evolution was 15 years for CD and 11 years for HS. CD had been previously confirmed by colonoscopy and biopsies in all patients. CD was active in all patients; 2 patients were under adalimumab, one patient had stopped ustekinumab 4 months before starting targeted strategy and 9 patients had stopped all treatments for CD for several years. Patients had been treated with a median of 4 immunosuppressive agents for their CD. 2 patients were in Hurley stage 3 HS at inclusion, 6 were in Hurley 2, 2 were in Hurley 1 and 1 had no active lesions. Treatments used were cotrimoxazole only in one case, ceftriaxone and metronidazole followed by rifampicin, moxifloxacin and metronidazole in one case and ertapenem in 9 cases. 8/11 patients had an improvement in their diarrhea, 1 patient was stable and 2 patients worsened. The median number of stools diminished from 6 to 2 after HS treatment. After HS induction treatment, all HS patients were in remission or much improved, depending on the severity of HS, except for one worsened patient who previously had many antibiotics for CD.

Unexpectedly, while using targeted antibiotherapy for HS, we observed a decrease in diarrhea in 8/11 patients under this strategy, without any modification in CD treatment. There are similarities in CD and HS: Nod 2 function is bacterial killing of gut commensal flora, suggesting an important role of microbiome in CD, while microbiome has been recently involved in HS. Tobacco is a risk factor associated with both diseases. Fibrosis can be observed in both affections, forming circumferential stenosis in CD and hypertrophic scars in HS. Antibiotics have been often used in HS with various results, but rarely in associations, while there are few reports of combinations of antibiotics in CD showing efficacy. Concomitant improvement of CD and HS with targeted antibiotics questions a similar mechanism of action, possibly via manipulating commensal flora.

Targeted antibiotherapy may improve CD symptoms in HS patients. These results need to be confirmed by a prospective study with close attention to antibio-resistance.

094 PS33 - poster session | HS and ankylosing spondylitis, resulting from secukinumab in the control of the two entities

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HS is a serious and debilitating disease that affects 0.41% of the Brazilian population. According to international treatment recommendations, the use of antibiotics for 10 weeks, such as cyclins, and immunosuppressants such as adalimumab would be indicated as treatment options.

We report a case of a 32 year old woman with previous diagnosis of ankylosing spondylitis who developed HS after bariatric surgery 10 years ago. Spondylitis was treated with methotrexate, sulfasalazine, infliximab, etanercept and adalimumab, with partial response. However HS remained active, Hurley III, requiring frequent use of antibiotics, systemic and intralesional corticosteroids and surgical excisions. For better control of spondylitis, secukinumab was introduced two years ago. There was total control of the back pain. HS lesions resolved soon after the induction period and no new lesions emerged. Exactly during this period, intense erythematous lesions appeared on the scalp, with pruritus, erythematous lesions in the axillary folds and inframammary region. Psoriasiform and eczematous lesions appeared in the trunk and limbs. Mycological examination and cultures were negative. Histological examination confirmed psoriasiform dermatitis. The diagnosis of paradoxal psoriasis was considered. Methotrexate 20 mg/week was introduced orally, as well as topicals such as calcipotriol, betamethasone, clobetasol and tacrolimus. After six months, there was almost complete control of the lesions, although some lesions on the scalp were more relapsing. HS is a chronic immune-mediated disease and the control of inflammation is a challenge. The use of adalimumab is effective, with more than 50% of patients achieving HiSCR in 12 weeks of treatment. However, many patients are not good responders and require therapeutic associations or biological exchange to control disease activity. Anti-TNF-alpha is well established as effective in HS, and there are a number of cases reports showing benefits of anti-IL-1 and anti-IL-17. Preliminary studies show high levels of IL-17 in the lesions of HS and in the plasma of these patients, indicating a possible therapeutic target. Joint involvement is not infrequent in HS patients, with the axial column being the most affected. Investigating complaints of persistent joint pain of inflammatory rhythm in these patients and even subclinical involvement should be incorporated into the routine of the dermatologist. Finally, the development of psoriasis associated with the use of anti-TNF-alpha is known, but the anti-IL-17 as a paradoxical phenomenon is little reported and is more a particularity of this case. Conclusion: the case is peculiar due to the good action of anti-IL-17 in the control of HS and associated arthropathy and the occurrence of psoriasiform reaction during the use of this biological.

095 PS34 - poster session | Biologics in HS: where are we now?

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Pathogenesis of hidradenitis suppurativa (HS) is multifactorial, but dysregulation of proinflammatory and anti-inflammatory cytokines

seems to play a significant role. This provides a rationale for treatment of HS with biologic preparations. This study was undertaken to provide an overview of the available data on the effectiveness and safety profile of biological agents most commonly used in therapy of HS. There are presented both the preparations with the efficacy confirmed so far in large randomized trials, as well as new therapeutic solutions which were sometimes reported only in isolated cases in off-label therapy.

In total 85 papers were analyzed, mostly including case reports and case series, but a few bigger randomized controlled trials were also found. We performed an analysis of reported studies focusing particularly on TNF-a inhibitors which are most commonly used preparations. However, we provide also some proofs for the use of other biological agents.

When analyzing currently available data, the highest response rate (according to the criteria established for each study) was noticed for infliximab (IFX) comparing with adalimumab (ADA) and etanercept (ETA) (82%, 54% and 54%, respectively). However, it is important to emphasize that the highest quality of evidence was reported for ADA what makes the result of its efficacy most reliable. The studies for ADA were much bigger than for IFX and ETA and many more patients were also analyzed after ADA administration. Variable results have been reported with the use of other biological preparations. All biologics were found to be generally well tolerated with mostly mild adverse events.

Current systemic therapies for HS still remain a serious challenge. Biological preparations play an important role. Adalimumab, which is the only FDA/EMA-approved biologic agent in HS therapy, should be considered first in a group of studied preparations.

096 PS35 - poster session | The efficacy of modifying adalimumab 40 mg weekly to adalimumab 80 mg every two weeks for hidradenitis suppurativa: a multicenter retrospective cohort study

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The use of adalimumab 40 mg has been suggested as efficacious and is a treatment in the EDF evidence based guidelines. Currently, no evidence exists on the efficacy of adalimumab 40 mg weekly vs adalimumab 80 mg every two weeks regimens with validated outcomes like IHS410 and there are no studies on infectious events and adverse events with this two regimens.

We have performed a preliminary study with a primary objective of analyzing the short term efficacy of modifying Adalimumab 40 mg weekly to Adalimumab 80 mg every two weeks with the validated outcome IHS4, DLQI, HAD I PROS.

A secondary objective was reporting infectious events and adverse events on both dosifications. Exploratory analysis of correlation between outcomes and baseline characteristics have been performed in order to identify clinical parameters which predict clinical response efficacy and adverse events after modifying dosification Adalimumab regimens.

A retrospective multicenter cohort study. Member sites of HS and sites with national HS registries have been invited to participate. We have collected data at three Time points: Baseline starting point with Adalimumab 40 mg; Week of change to Adalimumab 80 mg every two weeks; Minimum of 12 weeks after dose modification We will present our results and conclusions on this preliminary study using real-life data from our everyday practice. We discuss our experience of modifying dosification regimens.

097 PS36 - poster session | Therapeutic management of 1428 subjects suffering from hidradenitis suppurativa: EpiVer study

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Epiver is a French multicenter study and one of the largest series of patients with Hidradenitis Suppurativa (HS) diagnosed by experts.

As part of a French network that studies HS (RésoVerneuil) we wanted to characterize the management of our patients with HS.

From March 2016 to December 2017 ResoVerneuil members anonymously recorded data on treatment. It was a collection of spontaneous statements of patients.

Antibiotic therapy was performed in 76.75% patients: multiple courses of treatment (49.37%), monotherapy (68.77%), dual therapy (28.99%), triple therapy (4.97%).25% of patients never benefited from antibiotic therapy and less than half had iterative treatment. Cures lasted on average 3.46 months. Surgical incisions and excisions were performed in 59.31% and in 41.6% patients, respectively. The average number of incisions was 3. The average number of excisions was 1.02. Miscellaneous therapies were: zinc (19.12%), isotretinoin(9.59%), depilatory laser (5.18%), non-conventional medicine(4.97%) and botulinum toxin (0.14%).

We may be surprised by the limited number of therapeutic procedures performed in a chronic condition such as HS. Therefore, we must consider a possible under-declaration by the patients, an insufficient medical and surgical care, the fatigue and a feeling of helplessness of our patients and maybe of the doctors too. These hypotheses are not mutually exclusive. 40% of patients did not receive any surgical treatment, whereas surgery is the only potentially curative treatment to date. Although the treatment of the HS is not

perfectly codified, HS therapy proposals probably deserve to be more studied and widely divulgated.

098 PS37 - poster session | Refractory inguinoscrotal suppurative hidradenitis successfully treated with surgery and adalimumab: the importance of the multidisciplinary approach

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Complex hidradenitis suppurativa cases are common in real clinical practice. In some cases, treatment in monotherapy with systemic drugs or surgery are not enough to control the disease. Our objective is to highlight the importance of the multidisciplinary approach in the treatment of hidradenitis suppurativa and to demonstrate how the combination of surgery and biological drugs may be able to control the disease.

We present the case of a 52-year-old man with severe inguinoscrotal suppurative hidradenitis (Hurley III) starting in adolescence. In 2012, he presented with penis and scrotum lymphedema, with a buried penis on the scrotal sac that caused urinary incontinence. Surgery was performed in a different hospital, with a subsequent worsening of the disease. He came two years later to our center and was intervened by the Plastic Surgery Department up to 7 occasions. A debridement of all tissue affected by hidradenitis in the scrotal, pubic and inguinal areas was performed with a coverage with free skin grafts of full and partial thickness. Despite of the clinical improvement, pain and intense suppuration persisted with new arising hidradenitis lesions in the intervened areas. He was evaluated in Dermatology in July 2016 and we decided to start antibiotic treatment combining rifampin and clindamycin 300 mg / 12 hours for 10 weeks. Clinical improvement was seen, but he presented immediate worsening after antibiotics suspension in association with gastrointestinal intolerance. On January 2017, we decided to start treatment with subcutaneous adalimumab.

After 18 months of adalimumab treatment, hidradenitis suppurativa is controlled, with a practically absence of pain and suppuration. Treatment is currently maintained on a regimen of 80 mg every two weeks and the patient has required a single surgery intervention to eliminate cicatricial tissue. No secondary effects have been observed in association with adalimumab therapy.

Collaboration between different specialties is essential in the management of patients with hidradenitis suppurativa. We present the case of a man with a complex inguinoscrotal suppurative hidradenitis, who has presented an adequate disease control after the combination of surgery and treatment with adalimumab.

099 PS38 - poster session | Multidisciplinary committee for surgical management of hidradenitis suppurativa: our experience

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Management of hidradenitis suppurativa (HS) requires an integrated medical and surgical management. We present our results, based on a three-year experience of a multidisciplinary medical-surgical committee.

In a descriptive cross-sectional study, we recorded 104 patients that were evaluated by the committee from September 2015 to July 2018. The committee is comprised by the departments of Dermatology, Plastic Surgery and Colorectal Surgery, and meetings are on a monthly basis.

Each patient was evaluated by the committee after presentation of the case by the dermatologist. If surgery was decided, it was assumed by one of the departments: 47.9% of the patients were operated by Plastic Surgery, 41.7% by Colorectal Surgery and 10.4% by the Department of Dermatology. Loss of patients with a surgical appointment was 18.3%.

Most patients had a moderate or severe HS (Hurley II, 35.8% and 46.3% Hurley III). The most frequently involved areas were axillae (51.9%) and groins (41.3%). Surgery was performed most frequently on axillae (38.3%) and gluteal areas (23.4%). Dermatologic ultrasound was used for clinical staging and follow-up in all patients, endo-anal ultrasonography in 18.3% of them, and magnetic resonance in 12.5%.

Deroofing was the most frequent surgical procedure (47% of patients that underwent an operation). In terms of surgical outcome, 68.7% of all patients achieved a satisfactory result, in 12.5% of them dehiscence of surgical site was reported, and a relapse was noted in 18.8%. Biologic therapy had been started before surgery in 18.3% of patients, and it was not interrupted for the surgery procedure. 12.5% of patients started biologic therapy after the surgery.

Multidisciplinary committees are a useful management tool, in order to better tailoring of the treatment and follow-up in these patients, particularly in severe HS.

100 PS39 - poster session | Severe hidradenitis suppurativa in sacrum multidisciplinary approach

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Hidradenitis suppurativa (HS) is a chronic inflammatory skin condition of the apocrine gland that causes significant quality of life impairment.

A 47-year-old man with no significant medical history was assessed by the Dermatology department during hospitalization. The patient referred 3-year history of multiple painful abscesses and exudative fistulous tracts in buttocks, sacrum and perianal region. He did not present lesions in other areas. It was classified as Hurley stage 3 and IHS4 of 27. The patient had never consulted before and no previous treatment had been started. Several microbial cultures were collected from the lesions and all the results were negative. The patient was evaluated by the Gastroenterology department and Chron's disease was ruled out.

Treatment with oral doxycycline and glucocorticoids was initiated. Afterwards, subcutaneous administration of adalimumab was started at a loading dose of 160 mg, followed by a dose of 80 mg at the second week of treatment and 40 mg on a weekly basis later. After one year of follow-up, the patient reported improvement in pain and a decrease in bleeding and exudates.

Adalimumab therapy was discontinued due to the finding of anemia, thrombocytosis and leukocytosis in a routine blood test, initially attributed to the treatment. After stopping treatment, the patient experienced a worsening of pain and a new lesion developed in his right thigh. A magnetic resonance was performed and showed inflammatory activity in multiple and complex fistulous tracts in his right buttock and intergluteal fold. Signs of myositis in the right buttock and sacrococcygeal osteomyelitis were found in the magnetic resonance.

Our patient underwent surgery by the General and Digestive Surgery department and the Plastic Surgery department after their evaluation. A temporary colostomy, a surgical debridement of all lesions and a meshed skin graft of the left thigh were performed.

At present, the patient does not have any active lesions. Hematology department ruled out the possibility of drug induced analytical abnormalities, so that restarting adalimumab is being considered currently.

We report the case of a patient with a localized form of HS of the buttocks and sacrum who has required multidisciplinary approach and complex surgical treatment with great morbidity.

101 PS40 - poster session | Efficacy and safety of biologic therapy in surgical management of hidradenitis suppurativa

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Adalimumab approval for the treatment of moderate and severe variants of hidradenitis suppurativa (HS) has become an important advance for this condition.

There is some controversy about the need of biologic therapy discontinuation in those cases where a surgery is indicated. This is justified by a possible increase of complications mainly related with infections.

Focusing on HS, there is a high interest in not withdrawing the biological therapy, given that the expected results of the surgery could be better. The hypothesis, that is widely accepted by the general and dermatologic surgeons, is that a patient with a well controlled inflammatory activity before, during and after the intervention will reduce immediate and late surgical complications, offering the best results in the HS management.

We present a retrospective study with the aim of assessing the efficacy and safety of TNF-α blockers in a case series of patients with moderate or severe HS with fasciocutaneous flap surgical indication to cover wide wounds related with the cutaneous disease that continued with the biological therapy along the process.

We included all the HS patients under biological therapy that were surgically treated with indication of complex flap to remove complex fistulas after achieving a HiSCR between 2016-2018 from our Department.

8 patients were included, 7 males and 1 female with ages between 34 and 65 (adalimumab 7 cases, infliximab 1 case).

Surgical locations were: axillae (4 cases), buttocks (2 cases), and inguinal and thigh in another patient.

All of them remained under treatment with antiTNF-α and maintained HiSCR after surgery, with a median follow up of 15 months. Only one patient that discontinued treatment with adalimumab because he required a stoma in order to avoid infection. After 3 months, because of the development of new flares, Adalimumab was reintroduced and HiSCR was achieved after 12 weeks.

During the short follow-up during the next 4 weeks after surgery, 3 patients had surgical wound dehiscence without infection

2 cases were managed with conservative measures to promote second intention healing and 1 case underwent a minor surgery. No long-term complications were detected.

Although it is necessary to individualize each case, along with our experience, we support the continuation of antiTNF-α therapy in those patients with HS who will undergo any cutaneous surgery.

102 PS41 - poster session | Effectiveness, safety and tolerability of drainage and punch-trocar assisted cryoinsufflation (cryopunch) in the treatment of inflammatory acute fluid collections in hidradenitis suppurativa patients

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Patients with hidradenitis suppurativa (HS) often go to emergency services or dermatology consultations with acute inflammatory fluid collections, usually clinically identified as abscesses. Therapeutic alternatives to these lesions are limited. The objective of this study is

to describe and explore the effectiveness, safety and tolerability of punch-trocar assisted cryoinsufflation (cryopunch) for the treatment of acute fluid collections of patients with HS.

Cryopunch was performed on 10 acute fluid collection of 10 consecutive patients with HS. Participants were evaluated at the beginning of the treatment, on week 4 and week 24. Effectiveness was assessed as complete response or no response according to clinical and ultrasonographic criteria. Pain numeric rating scale was used to assess tolerability, any potential adverse effects were noted. Participants were evaluated at the beginning of the treatment, on week 4 and week 24. Cryopunch was effective in the 70% (7/10) of the treated fluid collections at week 4, $P = 0.008$ and at week 24 $P = 0.008$. There was no recurrence of any of the lesions that responded to the treatment. An inverse association between the duration of the lesion and the effectiveness was observed. Median pain associated with cryopunch procedure was 2 (1.75-3.25). No adverse effects associated with the study intervention were observed.

We present a simple, inexpensive, safe, tolerable and effective procedure for the treatment of acute inflammatory fluid collections of patients with HS.

103 PS42 - poster session | Could be surgery helpful before treatment with adalimumab for severe hidradenitis?

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Hidradenitis suppurativa (HS) is an example of how scientific effort can turn upside down the management of a particular disease. In fact, in the last years, HS has focused lot of dermatologist attention. On this respect, it is generally accepted that the identification of a novel and effective treatment for HS, an anti-TNF monoclonal AB, named adalimumab, has been determinant to wake up medical scientific community about this commonly overlooked disease.

On this respect, clinical experience indicates that Adalimumab is effective in approximately 50% of HS patients.

It is our impression that adalimumab for very active (very inflamed lesions) may be less effective, resulting in a worse overall patient response to the treatment.

In this way, the surgical excision of very active lesions prior adalimumab treatment may be advisable.

Here we report two patients in whom the surgical excision of very active lesions resulted in a good response to adalimumab, while prior to the surgery these two patients did not respond to the treatment.

Patient 1: A 19-year-old woman with a Hurley III hidradenitis focused on groins. After two years treatment with topical antibiotics and oral clindamycin-rifampicin she underwent adalimumab treatment at approved doses for HS. After eight months of treatment it had to be stopped due to the absence of response (assessed by clinical- HiSCR score and by ecographic image comparison) and left groin was excised. Two months after adalimumab was reintroduced and this time, the patient suffered a clinical and ecographic improvement of the disease specially in the right groin (the not surgically excised one).

Patient 2: Similar to patient one, a 23-year-old woman with a severe HS in whom adalimumab treatment had to be interrupted due to lack of response. However after surgical excision of left groin, adalimumab treatment was reintroduced resulting in an excellent clinical and ecographic response.

In these two patients, surgical excision of very inflamed lesions prior adalimumab treatment, changed the clinical and ecographic response to this treatment, making them responders whereas they where no responders before.

Surgical excision of very active lesions may boost the effectiveness of the adalimumab treatment in patients with severe HS.

104 PS43 - poster session | Photodynamic therapy in the treatment of moderate-to-severe hidradenitis suppurativa: a report of 28 patients

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Hidradenitis suppurativa (HS) is a chronic and recurrent autoinflammatory disease which affects the apocrine glands-bearing areas. The treatment of HS continuous to be a challenging , and several therapeutic alternatives have been described. The use of photodynamic therapy (PDT) with 5-aminolevulic acid (5-ALA) or methyl ester (MAL) has been proposed as a treatment option for recalcitrant HS with variable results. However, all the studies that have been published to date are series of few patients, and each study was conducted differently.

To evaluate the effectiveness of PDT with 5-ALA clinical and ultrasound in the treatment of HS.

Retrospective study with 28 patients suffering from moderate to severe HS from two university hospitals in Spain treated with PDT and topic 5-ALA. The patients were evaluated before and 8 weeks after their last session. The parameters used to evaluate the effectiveness of the treatment were clinical and ultrasound. They were measured using Hurley, HS4, PGA the quality of life using the Dermatology Life Quality Index and a pain visual scale (EVA).

A total of 28 patients (15 men and 13 women) with a mean age of 46 years and HS Hurley II-III were included. All patients showed an improvement in DLQI and EVA 8 weeks after the end of treatment. In the ultrasound we observed a resolution of the lesions in 13 patients, partial resolution in 12 and poor in 3.

In our experience the use of PDT can play a role in the treatment of this difficult disease. The treatment was performed safely with transient local effects and showed a significant decrease of symptoms and life quality improvement. Results kept positive after 8 weeks after last session.

105 PS44 - poster session | VENUS Study: Vision of hidradenitis suppurativa by family doctors in Portugal

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Hidradenitis suppurativa (HS) is a chronic inflammatory skin disease with a diagnostic delay of 7 years. The family doctor is usually the first doctor the patient visits in the Portuguese health care system.
To describe the awareness and clinical approach of HS by family doctors in Portugal.
A cross-sectional study was conducted based on a questionnaire designed to assess general knowledge about HS.
We obtained 187 questionnaires answered by family doctors. The majority (97.3%) of physicians reported HS as the most likely diagnosis in a typical clinical history and 84% associated HS with chronicity. When questioned about HS and disability, only 43% of the respondents considered this association relevant. The majority (84%) admitted to refer these patients, mostly to a dermatologist (89.3%). Most clinicians considered that a purely surgical approach may not be curative (94.1%) but only 15% admitted that biologic drugs may be indicated. The responses of two groups in this sample (group A, n = 81, with <5 years of clinical practice, and group B, n = 103, with> 5 years) were compared. Group A associated HS more with antibiotics, smoking and obesity than group B, and this difference was statistically significant ($P < 0.001$). Regarding treatment, group A more frequently selected systemic antibiotics, immunomodulatory drugs and multidisciplinary approach as therapeutic possibilities than group B ($P < 0.05$).
Early diagnosis and appropriate treatment are crucial in the clinical approach to HS. This pilot study concludes that the Portuguese family doctors' awareness about HS can be improved. There are significant differences between the HS knowledge of doctors with <5 years of clinical practice and the most experienced physicians. Future educational actions in Portugal should focus not only on residents but also on family medicine specialists.

106 PS45 - poster session | Influence of the duration of hidradenitis suppurativa on the quality of life and pain in 1428 subjects: EpiVer study

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Epiver is a French multicenter study and one of the largest series of patients with Hidradenitis Suppurativa (HS) diagnosed by experts.
As part of a French network that studies HS (RésoVerneuil) we wanted to characterize the impact of HS duration on quality of life, pain and stage of the disease. The evolution of HS over time is poorly known.
From March 2016 to December 2017 ResoVerneuil members anonymously recorded the standardized examination data of their patients with HS. It was a collection of spontaneous statements of patients. Quality of life was assessed by the DLQI scale and pain using the Visual Analogue Scale (VAS).
The results were given by 5-year duration of evolution: <5 years, 5-10 years, 10-15 years, 15-20 years, 20-25 years, 25-30 years, 30 and more. Median DLQI was respectively: 10; 12; 14; 14; 14; 14; 13. Median VAS was respectively: 5; 6; 6; 6; 6; 7; 6; 6. Hurley Stage was: stage I: 50%, stage II: 39%, stage III: 11%; stage I: 42%, stage II: 42%, stage III: 16%; stage I: 45%, stage II: 40%, stage III: 15%; stage I: 47%, stage II: 35%, stage III: 18%; stage I: 38%, stage II: 42%, stage III: 20%; stage I: 40%, stage II: 39%, stage III: 21%.
The quality of life and pain were not influenced by the duration of the disease when considering an evolution more than 5 years. However, there was evidence of increasing stage III and

107 PS46 - poster session | Body mass index does not influence the quality of life and pain in 1428 subjects with hidradenitis suppurativa: EpiVer study

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Epiver is a French multicenter study and one of the largest series of patients with Hidradenitis Suppurativa (HS) diagnosed by experts.
As part of a French network that studies HS (RésoVerneuil) we wanted to characterize the impact of the body mass index (BMI) on quality of life and on pain of our patients with HS.
From March 2016 to December 2017 RésoVerneuil members anonymously recorded the standardized examination data of their patients with HS as part of the EpiVer study. It was a collection of spontaneous statements of patients. Quality of life was measured by DLQI and pain by Visual Analogue Scale(VAS).
Tending towards a normo-weight must be advisable based on proven notions of epidemiology concerning the cardiovascular and metabolic diseases. However, weight did not have a significant impact on pain and it had a very discreet effect on quality of life in patients with HS.
EpiVer data tend to show that correction of weight is not a therapeutic priority for the management of pain and of impaired quality of

life in patient with HS. Indeed, we did not find an aggravation of the DLQI nor an increased VAS in obese. The management of pain must precede that one of weight in patients with HS.

108 PS47 - poster session | Is umbilical inflammation a sign of an impending hidradenitis suppurativa flare? A case series

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Hidradenitis suppurativa (HS) is a disease that is still not fully understood. We present three cases of patients that in relation with HS flares experience a reddening of their umbilicus. A phenomenon that has not been described before in the literature.

A 41-year old woman with HS and a 10 + year medical history Crohn's disease, stoma and a known history of fistula of the vulva and perineum. She experienced reddening of her umbilicus as early as 2013. In 2015 while treated with adalimumab for her Crohn's disease her fistulas had progressed and she was referred to the Department of Dermatology, Zealand University Hospital where she was diagnosed with HS. The umbilical erythema initially subsided but recurred at intervals, according to the patient often in relation to HS flares. In late 2017, a hole appeared on her navel. A CT scan showed several fistulas but none to the umbilicus. In fall 2018 upon examination a small orificium could be found at the location of her umbilicus.

A 55-years old woman diagnosed with HS since 1982. She has no family history of HS, but of psoriasis (sister), and rheumatoid arthritis (both maternal grandparents). In her youth, she underwent more than 50 incisions/excisions of boils. At the age of 27, she underwent skin transplantation to both axilla and at the age of 31 of the skin of the genitofemoral folds and above the mons pubis. After her transplants, her disease was in remission for several years. During the last few years, her disease has flared again in her transplanted areas and she now has the feeling of underlying abscess formation during flares. Since the time of her original HS diagnosis, she experienced activity in her umbilicus before and during a HS flare as well as itching eczema in the area of her skin transplants. Presently she is stable on a treatment of topical resorcinol. At her last appointment, she presented with only erythema of her umbilicus and the sensation of nagging pain under her skin transplants.

A 33-years old woman with known disposition for HS, and for psoriasis (brother). Her skin presents HS primarily located to the vulva. Previously, when she had flares she experienced erythema of her umbilicus. She has also experienced intermittent suppuration of blood and pus per rectum. In 2017 she had a colonoscopy and MR scan performed without finding a diagnosis.

As our patients informed us that the reddening of the umbilicus preceded or co-occurred with HS flares, a local reaction related to HS could be suspected. On the other hand, our three patients all have either genetic disposition for psoriasis or showed symptoms relatable to inflammatory bowel disease both of which are associated with HS. It may be speculated that reddening and/or itching of the umbilical skin could present a prodromal sign of a HS flare. If this hypothesis is confirmed in larger series it may provide an opportunity to aggressively treat a flare before it truly begins.

109 PS48 - poster session | A case of severe hidradenitis suppurativa successfully treated with secukinumab

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Hidradenitis suppurativa (HS) is a chronic, inflammatory skin disease. According to European guidelines the treatment of HS depends on the severity of the disease and includes topic and systemic antibiotics, retinoids, laser, surgical excision and biologics. The only one biologic drug approved by the FDA to treat HS is adalimumab (TNF-alfa inhibitor). However, recent studies have shown increased interleukin (IL)-17 serum levels in patients with HS, that indicates a possible therapeutic target for other biological therapy. Here, we report a case of severe HS treated successfully with secukinumab (IL-17 inhibitor).

This is a case of a 25-year-old Caucasian man, who was suffering from HS for 7 years, presented with Hurley IIC stage localized on the chest and back as well as in the lower abdominal region, right and left groin. The first symptom was the onset of inflamed nodules in the left axilla in 2011. The patient was diagnosed with HS in 2013. Comorbidities of the patient were obesity Class II (BMI= 35.3 kg/m2) and depression. Neither smoking addiction nor family history of HS were recorded. The patient was previously unresponsive to oral antibiotics treatments, including amoxicillin, rifampicin with clindamycin, sulfamethoxazole, lymecycline and tetracycline. Moreover, acitretin and cyclosporine systemic administration also proved to be ineffective, only temporarily stopping the progression of the disease. Additionally, over the 7-year period of time, number of surgical interventions from incision and drainage to wide removal with skin grafts did not bring the expected benefits. Due to poor response to these treatments the secukinumab therapy was considered as possible therapeutic option. The patient received 8 subcutaneous injections 300 mg of secukinumab each over a five-month period between June and October 2018. Clinical efficacy was reported by reduction in HSSI score from 18 to 10 points, Sartorius HS score from 96 to 27 points, physician global assessment (PGA) score from very severe to mild and Hidradenitis Suppurativa Severity Score System (IHS4) from severe into mild. Pain visual analogue scale score was reduced from 7 to 0 points. In addition, a decline of 8 points (12 to 4) in DLQI score occurred what significantly improved patients'

quality of life. Overall, the therapy was well tolerated and no side effects were observed after Il-17 blockage treatment in this patient. IL-17 blockage therapy proved to be safe and effective. At the end of secukinumab treatment the patient experienced reduction of inflammatory skin lesions, absence of pain and improved quality of life. It appears that HS cases with a domination of inflammatory nodules may be successfully treated with biologics. Further observations are needed.

110 PS49 - poster session | Staphylococcus aureus carriage status in patients with hidradenitis suppurativa

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Hidradenitis suppurativa (HS) is a chronic, recurrent, auto-inflammatory disease that affects the pilosebacous unit in apocrine gland-bearing areas. Bacteria are thought to play a role in its obscure aetiopathogenesis. In addition, antibiotics are frequently used, as first-line management for HS. We sought to determine the carriage status of Staphylococcus aureus in patients with HS at different stages of the disease.

This was a prospective observational study. Consecutive patients attending the HS clinic in a tertiary referral University Hospital, were included. Patients were either newly diagnosed or under treatment for HS, but no one had received antibiotic therapy for any reason during the previous three months. Nasal and oropharyngeal sampling was obtained and specimens were tested for the presence of S. aureus. Swabs were first incolulated in thioglycollate enrichment broth and then streaked on both mannitol salt agar and 5% sheep blood agar plates. Identification was performed based on colonial morphology, Gram stain, catalase, latex agglutination testing for coagulase detection and finally confirmed by the use of VITEK 2 system (bioMerieux, Marcy-l'Étoile, France). Antibiotic susceptibility testing was performed using the VITEK 2 system. The detection of MRSA was performed by the VITEK2 cefoxitin screening test. Standard statistical tests, descriptive statistics tests, chi square and Pearson correlation were performed using IBM SPSS Statistics 25. The level of significance was set at a P value <0.05.

Twenty-seven adult patients with HS were studied. There were 20 females (74.1%) and 7 males (25.9%). The mean age was 37.35 ± 13.57, and the mean age at onset of disease was 25.64 ± 12.91. The mean duration of disease was 12.32 ± 8.63 years. Six (22.2%) of the patients were Hurley stage I, five (18.5%) were Hurley stage II, and fourteen (51.9%) were Hurley stage III. Nine (33.3%) of the patients were on treatment with adalimumab. The prevalence of S. aureus carriage was 26% (7/27). Among these seven patients, 3(42.9%) had MRSA strains, and four had MSSA strains. S. aureus colonization and MRSA carriage were not statistically correlated with age, age of onset, BMI, smoking, and clinical severity (Hurley stage). A statistically significant correlation was found for S. aureus carriage (P = 0.029) and male gender.

In our study, the S. aureus carriage status among patients with HS was 26% and 42.8% of them had methicillin resistant Staphylococcus aureus (MRSA). Male patients were more probable to have S. aureus colonization. These are preliminary results of an ongoing study. A larger number of patients is necessary to establish more solid conclusions.

111 PS50 - poster session | The microbiome of tunnels in hidradenitis suppurativa patients

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Hidradenitis Suppurativa (HS) is a chronic inflammatory skin disease defined by recurrent nodules, tunnels and scarring involving the intertriginous regions. Recent Next Generation Sequencing (NGS) studies suggest genera such as Prevotella spp., Peptoniphilus spp. and Porphyromonas spp. are associated to chronic - and early HS lesions. However, a systematic investigation of the bacterial microbiome in HS tunnels remains unexplored using NGS.

We aimed to investigate the bacterial composition of the luminal white gelatinous material found in HS tunnels using NGS.

An exploratory study of patients with diagnosis of HS (n = 32) with tunnels. The tunnels were present either in the groin (n = 17) or in the axilla (n = 15). During deroofing of the tunnels, a sterile E-swab was taken of the luminal gelatinous material. The samples were investigated using NGS targeting 16S ribosomal RNA.

The skin microbiome was characterized in 32 HS patients. Overall, 5 microbiome types were identified: Porphyromonas spp. (Type I), Corynebacterium spp., (Type II), Staphylococcus spp. (Type III), Prevotella spp. (Type IV) and Acinetobacter spp (Type V). Porphyromonas spp. (Type I) and Prevotella spp. (IV) were the most frequent genera found in the tunnels.

This study points to a potential association between the presence of certain anaerobic bacteria (Porphyromonas spp., Prevotella spp.) and HS tunnels. Interestingly, these genera have previously been described in early inflamed and suppurating HS lesions using NGS. Thus, it may be speculated that these two genera are associated to the pathogenesis in HS either as drivers or as biomarkers.

112 PS51 - poster session | 3D-SeboSkin model for human ex vivo studies of hidradenitis suppurativa/acne inversa

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Hidradenitis suppurativa/acne inversa (HS) is a chronic, recurrent skin disease of the hair follicle. It manifests with painful, deeply localized,

inflammatory skin lesions that occur in apocrine gland-rich areas of the skin, most commonly in the axillae and the inguinal and anogenital regions (Dessauer definition). Epidemiological studies have reported a prevalence of the disease in Europe of 1%. The central pathogenic event in HS is believed to be the occlusion of the upper parts of the hair follicle leading to a perifollicular lympho-histiocytic inflammation. However, the exact pathogenesis of HS still needs further research.

To develop a human HS model which could help to understand the pathophysiology of HS and to determine the action of therapeutic candidates the 3D-SeboSkin technology (Nikolakis G et al. Exp Dermatol 24:497-502, 2015) was applied to co-culture explant involved and uninvolved skin of HS patients in direct contact with SZ95 sebaceous gland cell cultures for 3 days. The data obtained was compared with skin specimens from the same patients cultured alone under similar conditions. Detection of differential expression of HS biomarkers detected in own previous work at the mRNA and protein level was performed by H&ampE staining and immunohistochemistry in a group of female patients (median age 42 years, range 38-48, n = 4). The study was approved by the Ethics Committee of the Charité Universitaetsmedizin Berlin.

Using the HS 3D-SeboSkin model the structural integrity of the epidermis could be better maintained in comparison with the skin specimens cultured alone. Moreover, the 3D-SeboSkin setting facilitated the reproduction of the differential expression and pattern of several HS biomarkers (S100A9, CK16, TMPRSS11D, SERPINB4) in epidermal and dermal tissue and the appendages.

In conclusion, we present the first, reproducible, human model for studying HS. Moreover, we confirm the evidence of a beneficial interaction between HS skin explants and human SZ95 sebocytes to conserve skin integrity ex vivo. These data corroborate the value of the presented HS 3D-SeboSkin model for further HS research.

113 PS52 - poster session | Upregulation of SERPINB3/B4 and S100A7-9 in hidradenitis suppurativa/acne inversa

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Hidradenitis suppurativa/acne inversa (HS) is a chronic inflammatory disease of the follicular portion of the folliculopilosebaceous unit mediated by many non-specific inflammatory cytokines including IL17, IL23, IL1 and TNF α. However, recently demonstrated signal pathways indicate the complexity of HS pathogenesis.

In this study we investigated the expression and localization of the transglutaminase inhibitory proteins (SERPINB3/B4) and the antimicrobial proteins (S100A7-A9) in HS by gene array analysis, qRT-PCR and immunohistochemistry.

Comparative analysis of gene expression in the lesional vs healthy appearing skin from microarray data of 8 affected patients and 4 healthy individuals showed 705 statistically significant upregulated and 482 downregulated genes (fold change >2, $P < 0.05$). In relation to the biopsies taken from control individuals, 994 genes were upregulated and 1046 downregulated. Overexpression of the keratocyte proteins SERPINB3/B4 and S100A7/A8/A9 was validated using qRT-PCR. SERPINB4 and S100A8/A9 were overexpressed in the lesional skin vs. non-lesional with average fold changes of 18.5, 23.0, and 28.3, respectively (n = 15). Immunohistochemistry staining showed an upregulation of these proteins in lesional skin, specifically in the epidermis as well as in the inner and outer sheath of hair follicle.

Our findings may implicate new pathways in the pathogenesis of HS, which are associated with interruption of essential cellular functions, e.g. protein cross linking at the terminal differentiation level of follicular keratinocytes.

114 PS53 - poster session | Impact of adalimumab treatment on 30 patients' quality of life

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Hidradenitis suppurativa/acne inversa is a chronic, inflammatory skin disease, characterized by recurrent, deep-seated nodules, abscesses, and/or sinus tracts with suppuration, leading to hypertrophic scarring. During the last years many treatments have been proposed to control the disease. Adalimumab is the only one that has been approved for the control of the severe stages. The objective of our study was to estimate the impact of adalimumab treatment on patients' quality of life.

The DLQI questionnaire was completed by 30 patients before commencing adalimumab treatment and 6 months later.

Of the 30 patients, 17 were male and 13 females. All patients were in Hurley stage III. The mean IHS4 score 17.18 before and 6.55 after treatment. The mean DLQI before treatment was 15.27 and six months later 7.47.

Adalimumab seems to be a treatment that impairs and benefits the quality of life of HS patients. We plan to expand our study in more patients and by using more psychometric standardized tests.

115 PS54 - poster session | Prevalence and associated factors of alexithymia in patients with hidradenitis suppurativa

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Hidradenitis suppurativa (HS) is a chronic inflammatory skin condition which negatively affects patients' mental health and quality of

life resulting in higher risk of developing various psychiatric and psychological conditions such as depression and anxiety.

Alexithymia, personality trait characterized by impairment in identifying, distinguishing and describing feelings to others, may be another psychological disorder associated with HS. Alexithymics demonstrate externally oriented thinking and deficiency in emotional communication. Such patients may tend to focus their attention externally to the skin. Therefore, the aim of the study was to assess the prevalence of alexithymia and its correlation to HS in patients suffering from this disease.

The study involved 100 patients diagnosed with HS, recruited between January 2017 and October 2018. The study group consists of 59 males and 41 females aged 18-59 (mean, 34 ± 12.2 years). Disease severity was distributed as follows, Hurley I - 27 %, Hurley II - 61%, Hurley III - 12%. Alexithymia was assessed by Twenty-Item Toronto Alexithymia Scale (TAS-20) questionnaire. The patients were classified into 3 groups based on their responses to the alexithymia measure, equal to or less than 51 points = non-alexithymia, equal to or greater than 61 points = alexithymia. Scores of 52 to 60 points = possible alexithymia. The alexithymic personality trait was identified while TAS-20 score was ≥51 points. HS severity was assessed with Hurley staging and Sartorius scale. Additionally, quality of life was evaluated with Dermatology Life Quality Index (DLQI).

Mean TAS-20 score was 53.7 ± 14.2 points (range 28-90 points). Alexithymia was observed in 32 % of patients with HS. The prevalence of alexithymia was higher in men than in women with 35,6% and 26,8% respectively. The alexithymic personality trait was identified in 58% of the subjects. The statistical analysis revealed significant correlation between alexithymia and DLQI ($P < 0.0001$; $R = 0.61$) and Sartorius score ($P < 0.0003$; $R = 0.36$). In addition, smoking patients presented statistically significantly higher TAS-20 scores compared to non-smoking subjects ($P < 0.0001$).

Our results demonstrate high prevalence of alexithymia in HS patients. It occurs more frequently among HS patients compared to the general population. It seems that alexithymia can be considered as another HS psychological comorbidity. Because of its strong correlation with DLQI, alexithymia has a negative impact on patients' quality of life. That is the reason why dermatologists should be encouraged to use the TAS-20 in clinical practice in order to identify alexithymics. Lack of early identification may lead to decreased compliance to actually follow the medical advice which results in bad health outcomes

116 PS55 - poster session | The burden of teledermatological treatment of hidradenitis suppurativa in the Faroe Islands

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Teledermatology is an ingenious idea within a field of medicine relying so strongly on morphologic appearance.(1) It can bring expert assessment of cases to under-served areas like rural Australia(2) Sub-Saharan Africa(3) and the Faroe Islands,(4). From the perspective of the specialist, teledermatology is frequently being proposed as a means of reducing not only burden of treatment but also cost.(1, 5) The relative burden of teledermatological treatment of hidradenitis suppurativa (HS) is unknown, but increased knowledge of HS's place in teledermatology may help form future decisions regarding this type of management.

All specialist dermatology services in the Faroe Islands (Pop. 49 864 (2017)) have since 2003 been provided by the Department of Dermatology, Zealand University Hospital through a mix of traditional (800/year) and teledermatological (Approx. 2400/year) consultations. We performed a retrospective review of the teledermatology database since 2003. Descriptive statistics and simple comparisons between diagnostic entities are provided.

In the period 2003-2018 10,713 Faroese were diagnosed by a dermatologist and managed by teledermatology across 803 different diagnoses. Descriptive statistics of the group in general and for the 10 most common diagnoses can be found in Table I. A total of 30 patients with HS were seen (rank 65 among diagnoses). Amongst the 100 most frequent diagnoses we calculated the consultations per patient (CPP) ratio (we chose the top 100 to avoiding rare diseases with only a single diagnosed patient that was seen multiple times) for an average of 1.433 teledermatological consultations. Table II shows the 10 diseases with the highest teledermatologic burden per patient.

As concluded previously(3) the practice of teledermatology is very advantageous for diseases such as acne and other diseases where the patient can carry out treatment at home and the dermatologist just needs to ascertain the effect.

Half the point of teledermatology is that it is supposed to be quick and easy for the dermatologist offering a means of reducing both burden of treatment and cost in our increasingly hectic clinics. Thus, teledermatology should be for simple diseases, where patients do not require multiple consultations. Unfortunately, as is apparent from our data, HS is not such a disease. This is most evident in regards to treatment. Without the physical presence of a dermatologist, restrictions are imposed on both examination and treatment. The extent of sinus tracts can be hard to completely evaluate from pictures alone, and patients with severe HS do not merely require simple drainage of boils, but rather an advanced combination of medical treatment and dermatologic surgery. In conclusion, teledermatology may have a place in the diagnosis of HS in rural areas but falls short in regards to treatment for severe cases, as is evident by the multiple CPP needed.

117 PS56 - poster session | Itch and pain influence on sleep in hidradenitis suppurativa patients: comparison to other chronic dermatoses

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Hidradenitis suppurativa (HS) is a chronic, inflammatory and debilitating skin disease of the hair follicle, associated with pain of

inflamed lesions in the apocrine gland-bearing areas of the body. Atopic dermatitis (AD) and plaque psoriasis (Ps) are also inflammatory, chronic and recurrent dermatoses. HS, AD and Ps, as well as accompanying them subjective symptoms (itch and pain), have great impact on patients' well-being. The study was undertaken to evaluate the influence of itch and pain on sleep in HS patients compared to AD and Ps patients.

The study group consisted of 108 HS patients (51 females, 57 males; mean age 36.3 ± 12.1 years) versus 100 AD patients (42 females, 58 males; mean age 39.2 ± 15.4 years) and 100 Ps patients (39 females, 61 males, mean age years 44.1 ± 15.8 years). The mean HS severity was assessed as 34.8 ± 32.1 points, 9.0 ± 4.4 points and 50/49/9, 33.6 ± 10.7 points and 13.5 ± 8.4 points according to HSS (Hidradenitis Suppurativa Score), HSSI (Hidradenitis Suppurativa Severity Index), Hurley I/II/III staging, SCORAD (SCORing Atopic Dermatitis) and PASI (Psoriasis Area Severity Index), respectively). Itch and pain intensity were evaluated with visual analogue scale (VAS). The sleep disturbances were estimated with Athens Insomnia Scale (AIS) and Pittsburgh Sleep Quality Index (PSQI). Moreover, the quality of life (QoL) was assessed by DLQI.

The mean itch and pain intensity within three last days was estimated as: HS: 4.1 ± 2.9 points and 4.9 ± 2.9 points, AD: 7.1 ± 2.7 points and 5.3 ± 2.9 points, and Ps: 6.6 ± 2.6 points and 6.5 ± 3.6 points, respectively. The mean AIS and PSQI scores were in HS: 5.4 ± 4.3 points and 6.5 ± 3.6 points, in AD: 10.5 ± 5.5 points and 8.3 ± 4.2 points and in Ps: 7.4 ± 5.2 points and 8.1 ± 4.8 points. Additionally, the AIS total scores were significantly different among HS compared to AD and Ps, ($P < 0.0001$) and ($P = 0.02$), respectively. The obtained results for PSQI total scores were significantly higher for AD compared to HS ($P = 0.02$). The average QoL was estimated as 13.0 ± 8.0 points, 16.4 ± 7.9 points and 12.8 ± 7.5 points, for HS, AD and Ps, respectively. The intensity of itch and pain significantly correlated with scores obtained by the AIS among HS patients ($r = 0.24$, $P = 0.03$, for both correlations). Furthermore, presence of itch and pain had a significant impact on insomnia frequency, but the pain was additionally a crucial factor responsible for poor sleep quality. The itch severity significantly correlated with scores obtained by AIS in AD patients ($r = 0.44$, $P < 0.0001$) and among Ps patients by AIS and PSQI scores ($r = 0.34$, $P < 0.001$ and $r = 0.59$, $P < 0.001$, respectively).

Itch has an important impact on insomnia and sleep quality in patients with chronic dermatoses, such as HS, AD and Ps. Moreover, pain is substantial related on insomnia and sleep quality among HS patients.

118 PS57 - poster session | Clinical and epidemiological features associated with pruritus and bad odor in patients with hidradenitis suppurativa

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Patients with hidradenitis suppurativa usually suffer from subjective symptoms like pruritus and bad odor. The objective of this study is to explore which clinical and epidemiological features are potentially associated with these symptoms and their impact on quality of life.

Cross-sectional study. Pruritus and bad odor were assessed by means of the Visual Analogue Scale (VAS). Clinical and epidemiological features were assessed through clinical interview, physical examination and cutaneous ultrasonography. Dermatology Life Quality Index (DLQI) was used to assess quality of life.

One hundred thirty four patients were enrolled in the study. Pruritus intensity was positively associated with female sex, submammary, inguinal lesions, number of areas involved and acneiform phenotype. There was no association between disease severity Hurley stage, IHS4, number of lesions and pruritus intensity. Bad odor was associated with abdominal and axial lesions, increasing weight, body mass index and disease duration, IHS4, Hurley stage, number of abscesses, fistulas and areas involved. Both pruritus and bad odor were associated with increasing pain, suppuration and worse quality of life.

Our study shows that pruritus and bad odor have a negative effect on quality of life. Itch is probably not associated with disease severity but some disease locations appear to favor this symptom. On the other hand, bad odor correlates with disease severity and number of suppurative lesions. Increasing weight is also associated with higher levels of bad odor.