Introduction
Severe hyperandrogenism is a warning sign in young women. Differential diagnosis includes neoplastic, non-neoplastic and iatrogenic causes. The association of hyperandrogenism and congenital or acquired portosystemic shunt (PSS) has been rarely described, with its pathophysiology being unclear.
Case 1
A 22 year-old woman with diagnosis of autoimmune hepatitis, portal hypertension and PSS since 15 years old, presented with a 4 years history of oligomenorrhea and hirsutism [Ferriman-Gallwey (FG) score 12]. Examination revealed BMI 30.7 Kg/m^2 and virilization signs. Total testosterone (TT) was 3.60 ng/ml (0.1–0.56) and androstenedione 12.4 ng/ml (0.5–4.7). 17OHP, S-DHEA and SHBG were normal. Serum glucose 83 mg/dl, insulin 20 uUI/ml (1.9–23), HOMA-IR 0.7. CT scan excluded adrenal lesions. Ultrasound showed "ovaries with 56 and 58 mm and multiple peripheral follicles". In view of the severe hyperandrogenism of probable ovarian etiology, catheterization of the ovarian veins was performed, excluding ovarian lateralization or central/peripheral gradient of androgen levels. The patient completed 17 months of goserrelin acetate with slight improvement in hyperandrogenism (TT 1.88 ng/ml, androstenedione 8.58 ng/ml). Due to poor therapeutic adherence and necessary contraception, an ethonogestrel implant was applied.
Case 2
A 18 year old woman, with portal hypertension due to portal vein agenesis diagnosed at 4 years old, presented with menstrual irregularities and hirsutism (FG score 10) since menarche at 14. She had a BMI 28.9 Kg/m^2 and no signs of virilization. TT was 2.12 ng/ml and androstenedione 5.27 ng/ml. 17OHP, S-DHEA and SHBG were normal. Serum glucose was 71 mg/dl and insulin 12.57 uUi/ml, HOMA-IR 2.2. On ultrasound, "right ovary (30.4 × 26.4 mm) with functional cyst and multifollicular left ovary (31.4 x16 mm)" stood out. Abdominal-pelvic MRI confirmed portal vein agenesis and excluded ovarian and adrenal lesions. In view of the mild clinical symptoms and the need for contraception, a levonorgestrel IUD was inserted.
Conclusion
PSS can cause hyperinsulinemia by direct passage of secreted insulin to the systemic circulation and consequent insulin resistance by downregulation mechanisms. This is a possible explanation for the hyperandrogenism associated with PSS, similarly to what happens in polycystic ovary syndrome. Decreased hepatic clearance of androgens, in particular testosterone, may be an additional factor. Treatment depends on age, degree of virilization and pregnancy goal which justified the different approach in these patients.

DOI: 10.1530/endoabs.73.AEP582

AEP583

Low intelligent quotient (IQ) in patients with Klinefelter Syndrome are associated with impaired quality of life: A systematic review with meta-analysis

Brien Mehmet & Sofia Llahana
City, University of London, School of Health Sciences, London, United Kingdom

Objective
This was a systematic review with meta-analysis aiming to identify if patients with Klinefelter syndrome (KS) had a reduced full scale intelligent quotient (IQ) when compared to controls. Reduced IQ is shown to have a negative multifaceted effect on individuals' Quality of Life (QoL), having been shown as a *predictor of future success, increased criminal behaviour, post-traumatic stress disorder (PTSD), lower academic achievements and increased prosocial deficits*. Assessment of patients' IQ can support clinicians in delivering patient care intervention which can address individualised QoL deficits and patients' unmet needs. This is particularly relevant and crucial in achieving holistic nursing care to intervention.
Design
Meta-analysis was completed in Review manager 5.4, using continuous data and running an inverse variance random-effects model, using Std. mean difference for the effect measure, a forest plot was created. This analysed the results on full scale IQ from all studies that used both controls, KS participants and a validated measuring tool to record IQ. Seven studies in total were appropriate to be combined for meta-analysis. The seven studies included were extracted from the initial systematic review analysing factors that can influence QoL in patients with KS.
Data sources
Medline, Cochrane, Embase, Psychinfo, CINAHL, BASE and grey search from the reference lists of key publications.
Eligibility criteria
RCT's, Cohort studies, cross sectional studies and Epidemiology studies involving patients with KS and reporting on QoL parameters. Both adult and paediatric participants were included.
Results
The results from the meta-analysis suggest association with a lower full-scale IQ and a KS diagnosis. There is strong significant difference between patients with KS and Controls, significant P and Z values mean the probability of achieving the results by chance are lowered and the null hypothesis can be rejected. The statistical differences identified suggest a negative association between full scale IQ and KS when compared to controls, suggesting lower full-scale IQ can be associated with KS.
Conclusions
Significant Z & *P* values (Z=8.10, *P* <0.00001) indicate that men with KS have a significantly lower IQ than healthy controls which has a negative impact on patients' QoL. Currently there are no validated scales to measure QoL parameters, including IQ, for patients with KS. Future research is needed to develop a KS-specific scale for use in clinical practice to identify patients' deficits in QoL parameters and plan appropriate care management plans to improve patients' QoL.
Prospero registration number - CRD4202017343.

DOI: 10.1530/endoabs.73.AEP583

AEP584

Primary ovarian failure: descriptive study of 50 cases

Wajdi Safi[1], Mouna Elleuch[2], Dhouha BenSalah[1], Hamdi Frikha[1], Nadia Charfi[1], Nabila Rekik[1], Fatma Mnif[1], Mouna Mnif Feki[1], Faten Hadj Kacem[1], Mohamed Abid[1]
[1]Hedi Chaker hospital, Sfax; Tunisia, Sfax, Tunisia, Department of Endocrinology, Sfax, Tunisia

Introduction
Primary ovarian failure currently represents an increasingly frequent cause for consultation in endocrinology, from 4 to 18 % of the causes of primary amenorrhea and 10 to 28 % of the causes of secondary amenorrhea.
Objectives
In this context, we report a retrospective study of 50 patients followed between 2000 and 2020 for early menopause, in order to assess the etiologies and risk factors favouring this pathology as well as its subsequent impact.
Results
The average age of our patients was 31 years with extremes ranging from 10 years to 40 years. A family history of early menopause was noted in only 6% of women. The reason for consultation was secondary amenorrhea in 52% of cases followed by primary amenorrhea in 32%. In our study we found that the majority of women are nulliparous, in 68% of cases. The diagnosis was confirmed by an average FSH level of 87.25 mIU/l, LH at 28.45 mIU/l, the average E2 value was 2.4 pg/ml and the average prolactin value was 29.02ng/ml The study showed that the main aetiology was premature autoimmune ovarian failure in 58% of cases, a congenital cause in 24% of cases, and chemotherapy and radiotherapy are involved in 6% of cases. All the women in our survey presented complications following their menopause, of which 70% presented short-term complications, mainly sleep disorders, hot flashes and urinary genital disorders, 20% of the women presented metabolic complications at medium term and long term and 48% of women presented mainly cardiovascular complications.
Conclusion
Hormone replacement therapy is necessary to avoid the increased risk of all these complications, especially osteoporosis and cardiovascular complications. If you want to become pregnant, in vitro fertilization with oocyte donation is currently the most effective technique

DOI: 10.1530/endoabs.73.AEP584

AEP585

Long-term psychosocial effects of gender affirming hormone therapy on transgender men

Emre Durcan[1], Basak Ecem Bircan[1], Selver Yaylamaz[1], Sabriye Sibel Taze[1], Hande Mefkure Ozkaya[1], Senol Turan[2] & Pinar Kadioglu[1]
[1]Istanbul University-Cerrahpasa, Cerrahpasa Medical School, Department of Internal Medicine, Division of Endocrinology and Metabolism, Turkey; [2]Istanbul University-Cerrahpasa, Cerrahpasa Medical School, Department of Psychiatry, Turkey