

# Hyperandrogenism in a 12-year old girl with a congenital porto-systemic shunt and congenital hepatic fibrosis

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## Background:

Within the last 15 years single case-reports of hyperandrogenism in female patients with congenital porto-systemic shunts were described in literature. The mechanism of such coincidence is unknown but the role of hyperinsulinism, impaired androgens liver metabolism due to escape of some part of hormones via shunt from portal to systemic circulation are mentioned.

## Case presentation:

A 12-year old girl with increasing hyperandrogenism was admitted to Endocrinology Clinic.

### On examination:

- hirsutism (13-14 pts in Ferriman-Gallwey scale),
- severe acne,
- clitoromegaly
- low-pitched voice

### On anamnesis:

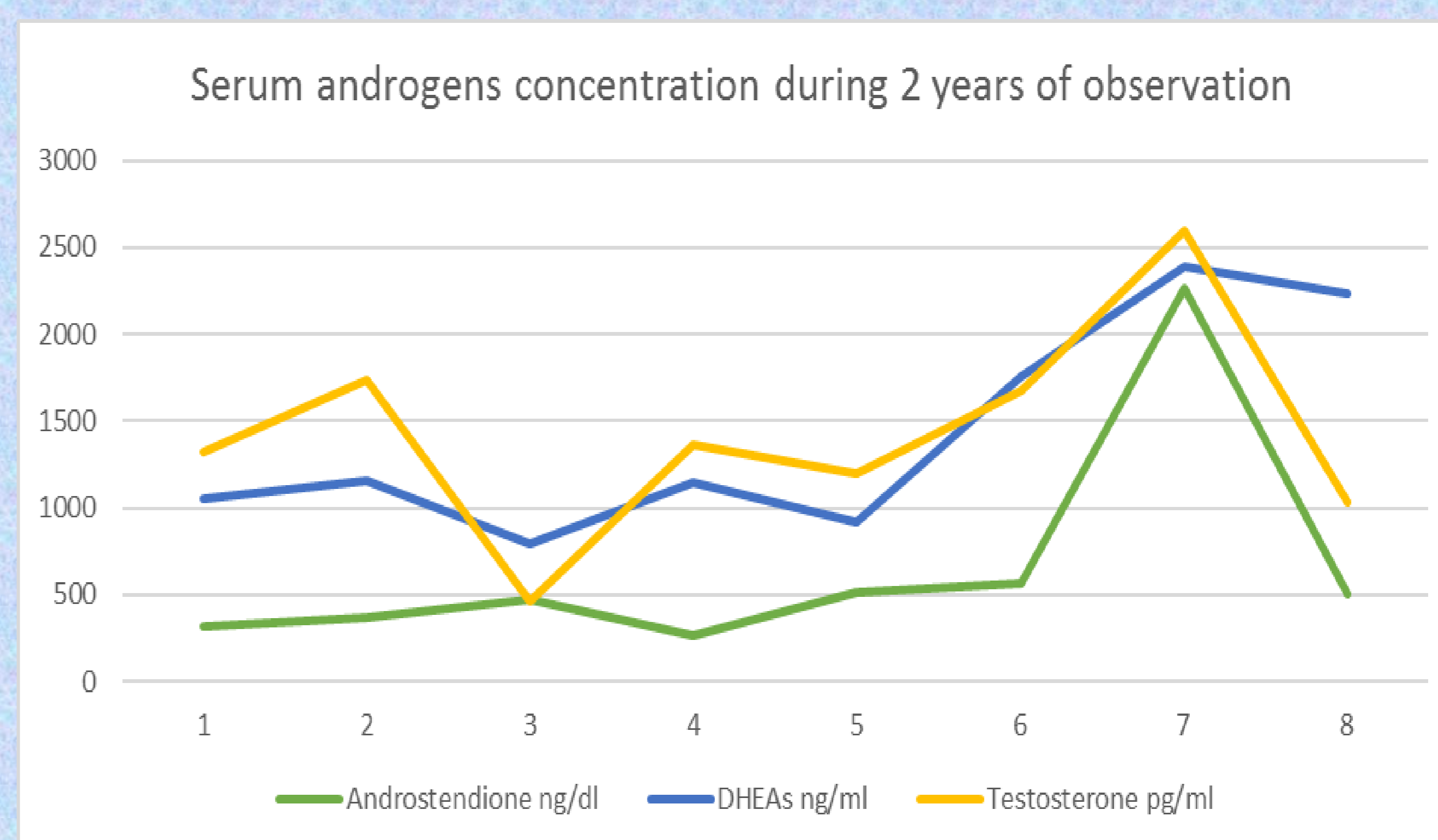
- congenital liver fibrosis
- mild portal hypertension
- 11y.surgery because of pancreas tumor (solid pseudopapillary tumor-Gruber-Franz Tumor).

### On laboratory tests:

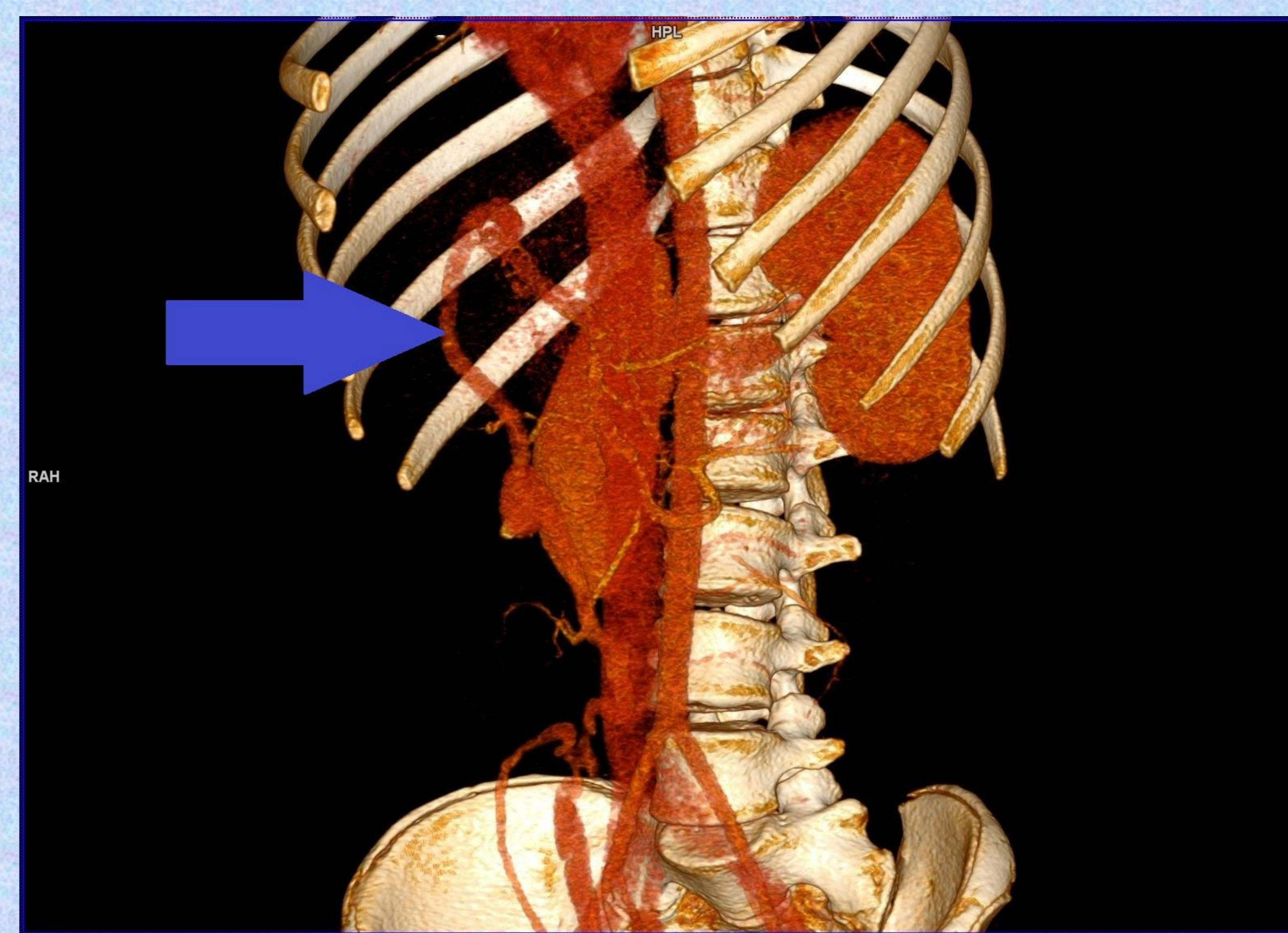
- high testosterone –max.2402 pg/ml (n < 950 pg/ml)
- androstenedione- max. 622 ng/dl (n < 470 ng/dl),
- dehydroepiandrosterone sulfate - within normal range
- steroid urinary profile - augmented androgen's metabolites
- long dexamethasone suppression test-
  - adrenal androgens and cortisol fully suppressed
  - partial testosterone and androstenedione suppression
 This suggested an ovarian contribution in androgen overproduction.
- OGTT: hyperinsulinemia (0' 24-40μIU/mL, 120' 120-210 μIU/mL) and impaired glucose tolerance.
- elevated serum ammonia concentration-120-180 μg/dl (normal range 20-80 μg/dl).

### Diagnostic imaging:

- brain and abdominal MRI - no changes
- abdominal angio-CT examination - porto-systemic shunt - persistent umbilical vein connecting portal with femoral vein.



Serum Insulin concentration in OGTT μIU/ml		
0'	60'	120'
40	239	144
37,4	189	210
24	128	130



Fot.1 Persistent umbilical vein

## Conclusions

Taking under consideration clinical and diagnostic findings, absence of hormonally active lesion - in our opinion hyperandrogenism in this case may be related with congenital porto-systemic shunt, similarly to cases previously described in literature. Mechanism of fluctuation of androgen's concentration in our patient remains unclear.

### References:

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3. T. Timpanaro et al. Congenital Portosystemic Shunt: Our Experience, Case Reports in Paediatrics Volume 2015, Article ID 691618
4. H. Guo-Hua et al. Insight into congenital absence of the portal vein: Is it rare? World J Gastroenterol 2008 October 21; 14(39): 5969-5979