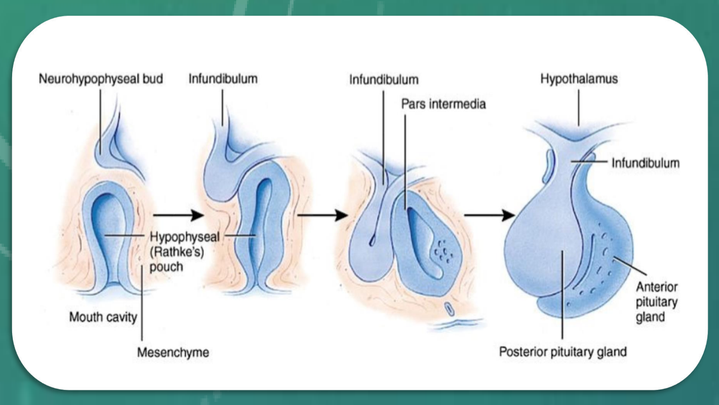
Pituitary stalk interruption syndrome (PSIS) is not a rare cause of the congenital hypopituitarism

Erdal Eren*, Zeynep Yazıcı**, Özgecan Demirbaş*, Nadide Başak Gülleroğlu**, Ömer Tarım*
* Uludag University, Faculty of Medicine, Department of Pediatric Endocrinology, Bursa, TURKEY
**Uludag University, Faculty of Medicine, Department of Pediatric Radiology, Bursa, TURKEY

Introduction and objective: Pituitary hypoplasia, empty sella syndrome, and ectopic neurohypophysis are common causes of pituitary MRI of the patient with congenital hypopituitarism (CH). We aimed to search clinical and radiological examination of the patient with CH.



Methods: We evaluated age, diagnosis, laboratory evaluation, hormone deficiencies, accompanying diseases, and MR images of the patients with multiple pituitary insufficiency.

Results: Of the 50 cases, 54 % (n = 27) were female with mean age of 6.43 ± 5.15 years, and 7.09 ± 5.21 years for admission and diagnosis, respectively ($p<0.05$). The reasons for referral were as follows; short stature (n = 36), jaundice and/or cholestasis (n = 5), hypoglycemia (n = 4), thyroid test abnormalities (n = 4), menstrual problems (n = 3), polyuria/enuresis (n = 2), cyanosis (n = 2), micropenis (n = 1), constipation (n = 1) and weight gain (n = 1) (Figure 1). The height, weight and BMI SDS were -3.18, -1.93 and -0.03, respectively (Figure 2). Hormonal evaluation revealed growth hormone deficiency (peak growth hormone response 0.84 ng / ml) in 48 cases, hypothyroidism (fT4 0.59 ng / dl, TSH 2.78 mIU / ml) in 40 cases, adrenal insufficiency (cortisol 2.8 mcg / dl, 14.2 pg / ml), sex hormone deficiency (LH 0.98 mIU / ml, E2 16.5 pg / ml, t. testosterone 0.86 ng / ml) in 25 cases. More than half of the cases with growth hormone deficiencies had very low IGF levels (<25 ng / ml). The third or fourth hormone deficiency appeared later in the 17 cases (34 %). **MR imaging showed PSIS (54 %) in 27 cases, anterior pituitary hypoplasia in 9 cases (18 %), partial empty sella in 6 cases (6 %), and normal pituitary defect in 11 cases (22 %).** When the patients were divided into two groups as having PSIS and not, the prolactin level was significantly higher (44.97 vs 11.9 ng / ml, $p <0.005$) and the st4 level was significantly lower (0.58 versus 0.60 ng / dl, $p = 0.019$) in the PSIS group (Figure 3).

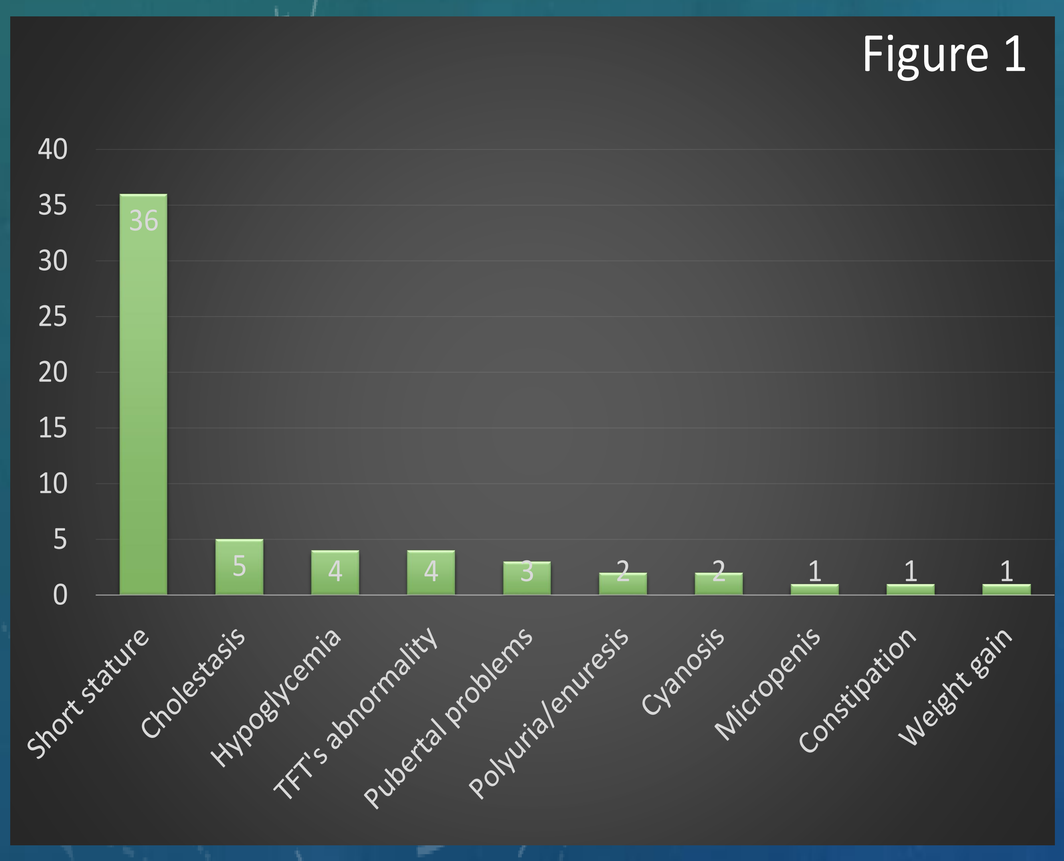


Figure 3

	PSIS (+)	PSIS (-)	p
Prolactin (ng/ml)	44.77	11.9	<0.005
sT4 (ng/ml)	0.58	0.60	0.019

Conclusion: Pituitary stalk interruption syndrome (PSIS) is a radiological diagnosis characterized by absent or thinning of the pituitary stalk, anterior hypoplasic pituitary, and ectopic posterior lobe. PSIS may be associated with midline anomalies and extrapituitary anomalies. The striking feature of the study was to diagnose PSIS more than expected with a significantly higher prolactin level. Increased prolactin may help to diagnose for PSIS. The diagnosis of PSIS that genetic cause is still unclear must be kept in the patient with hypopituitarism. Therefore, presence of stalk should be evaluated carefully in cranial imaging especially if there is an apparent ectopic neurohypophysis.

57ESPE
320--P2
Pituitary, neuroendocrinology and puberty
Erdal Eren
Poster presented at: 57th ESPE 2018 Meeting ATHENS GREECE 27-29 September 2018
PosterSessionOnline.com