

Pituitary Stalk Interruption Syndrome in Pediatrics: A Triad to Remember Forever

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Clinical Image

Pituitary stalk interruption syndrome (PSIS) is a very rare congenital syndrome, characterised by a triad combining: an absent or hypoplastic anterior pituitary gland, thin or absent infundibulum, and ectopic posterior pituitary location. It was first reported in 1987 by Fujisawa et al. in a patient with pituitary dwarfism and its reported incidence is to this date is only 0.5 in 1 million births [1]. Through this image article, we present the case of a 13-years-old girl who suffered from short stature and dyslexia. She was diagnosed with pituitary stalk interruption syndrome following an MRI in our department.

Clinically, patients usually present with deficiency of growth hormones which results in delayed growth. We could also see the deficiency progressing to other pituitary hormones with eventually the development of pan hypopituitarism with preservation of posterior pituitary function. Later in childhood, there could also be some learning difficulties or seizures [2]. We should note that the etiology of this syndrome is not yet perfectly established with some reports suggesting it may be the consequence of a defective migration of the pituitary gland during intrauterine life or ischaemia. The latter will then result to reorganisation of infundibular axons and thus develop an ectopic posterior pituitary.

As for imaging, we admit that MRI is the only modality adequately and perfectly able to identify the three features of the PSIS, which include [1-3]:

- Hypoplastic or absent anterior pituitary gland: the mean height of the entire pituitary gland varies with the age range. In pediatrics we admit a normal height of 5,6 +/- 1,6mm for infants between 1 and 10 years old, and a height of 6,3 +/- 1,8mm between 10 and 20 years old [4].

- Thin (better seen after Gadolinium administration) or totally absent infundibulum:
- Ectopic posterior pituitary: best seen on sagittal midline T1 images as high T1 signal measuring approximately 3 to 8mm tissue nodule at the median eminence (floor of the third ventricle), combined with an absent posterior pituitary bright spot.

Once the diagnosis is made, we may start the patient on hormonal replacement (Figure 1).

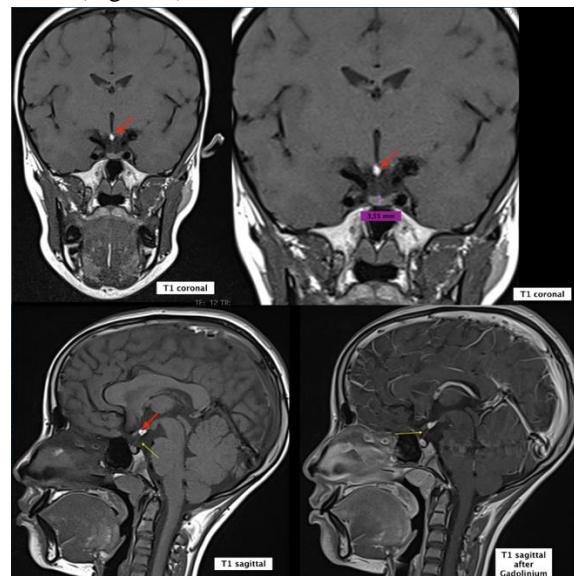


Figure 1: Our patient's T1 WI coronal and sagittal before and after Gadolinium administration demonstrating the triad of PSIS: high signal T1 nodular tissue in the floor of the third ventricle (red arrow) signifying ectopic posterior pituitary, very thin infundibulum better seen after Gado (yellow arrow) and hypoplastic anterior pituitary gland (in purple).

Received date: 20 April 2023; Accepted date: 24 April 2023; Published date: 27 April 2023

Citation: El Harras Y, Choayb S, Guennouni A, Allali N, Chat L, El Haddad S (2023) Pituitary Stalk Interruption Syndrome in Pediatrics: A Triad to Remember Forever. SunText Rev Case Rep Image 4(3): 177.

DOI: <https://doi.org/10.51737/2766-4589.2023.077>

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Conclusion

It is very important for radiologists to know the triad of the PSIS since MRI is the only imaging modality capable of diagnosing this abnormality and thus prevent preventing adverse effects on long-term growth and development.

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