Bartter syndrome complicated with growth hormone deficiency due to a suprasellar arachnoid cyst

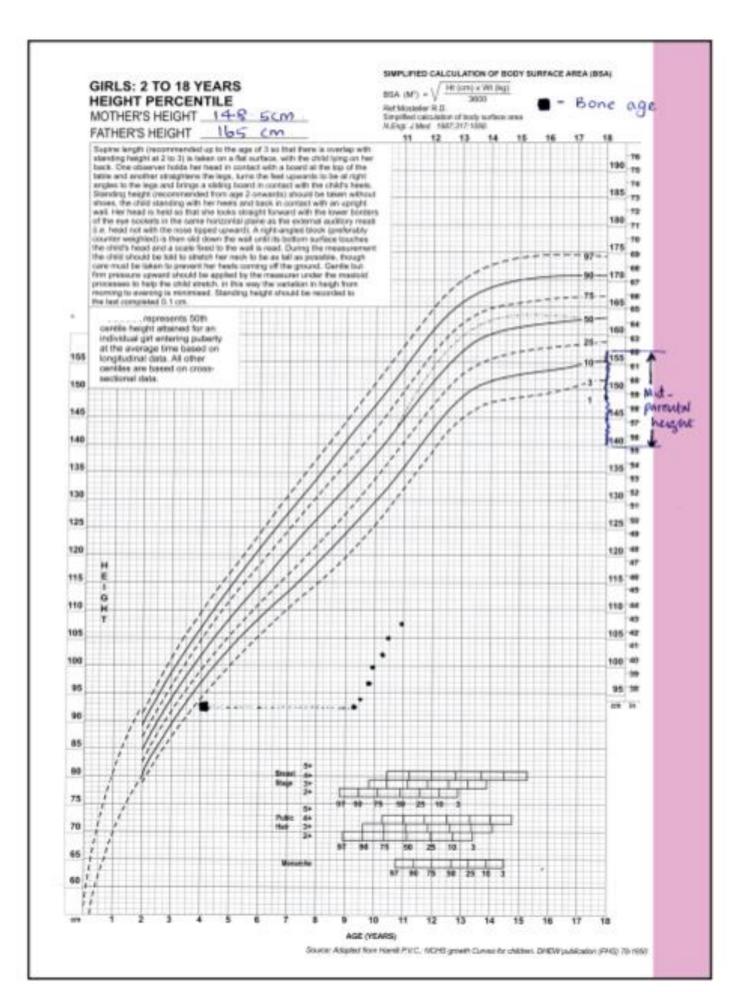
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Bartter syndrome affects about 1 per million people worldwide

Bartter syndrome (BS) is a rare genetic renal tubular disorder characterized by hypokalemia, salt-wasting and metabolic alkalosis. Polyuria, polydipsia, hypokalemia and salt loss are responsible for the growth retardation seen in BS. Persistent growth failure despite optimizing medical therapy may be due to growth hormone (GH) deficiency.

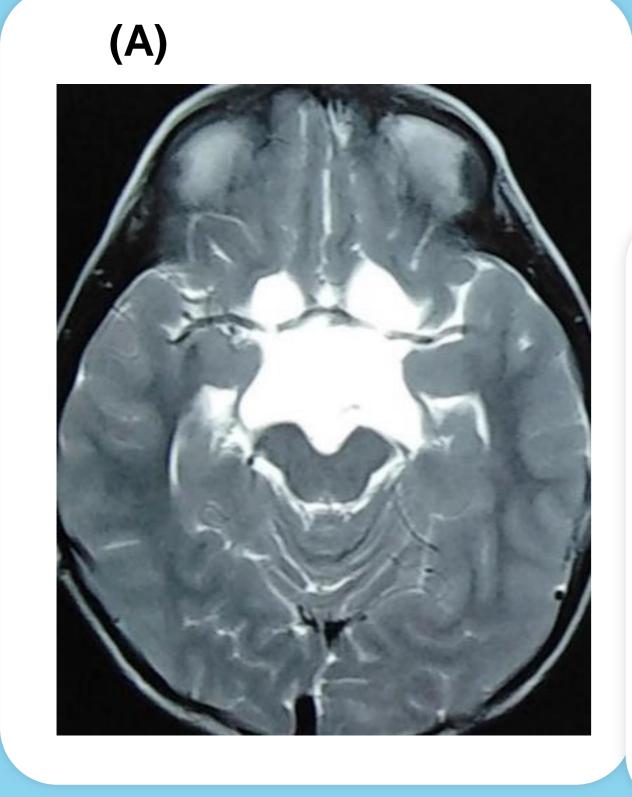
Diagnosing growth hormone deficiency

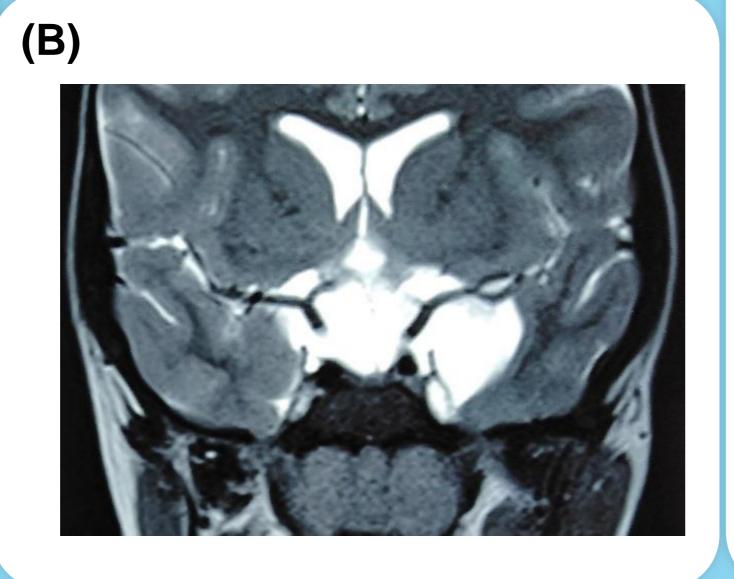
- A 9-year-old girl diagnosed with BS was referred for evaluation of severe short stature (92.5 cm, -7.7 SD).
- Her medical therapy consisted of indomethacin, salt and potassium supplements.
- Her bone age was 4 years and glucagon-stimulated growth hormone (GH) level showed a peak of 2.9 ng/dL. Reference range: <3 deficiency;
 3-7 partial deficiency; >7 normal
- Thyroid hormone and cortisol levels were normal.
- Magnetic resonance imaging (MRI) showed radiological features of a suprasellar arachnoid cyst.
- She showed excellent response to recombinant GH therapy (rGH). The absence of obstructive symptoms did not warrant a neurosurgical intervention.

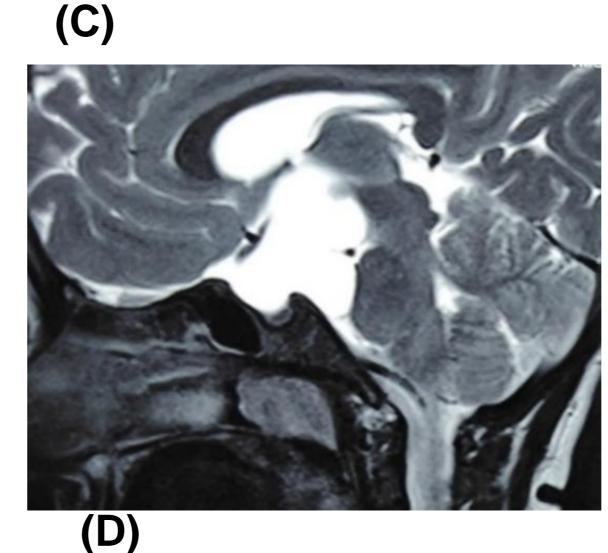


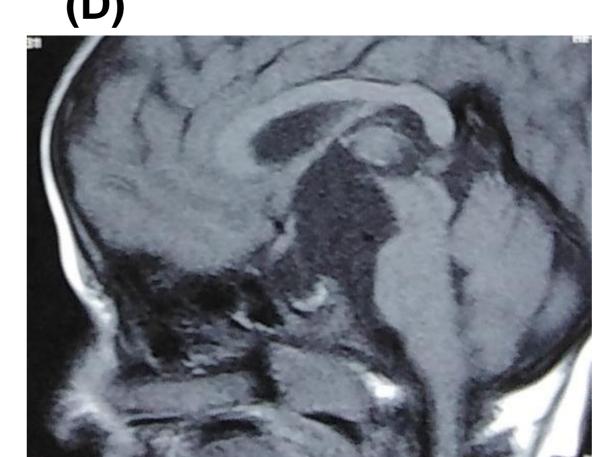
Significant growth spurt following rGH

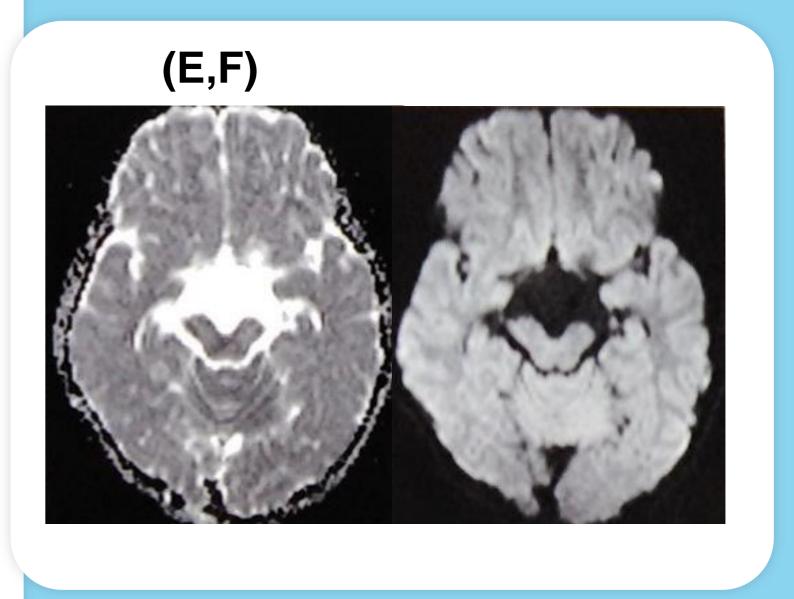
MRI Brain T2w axial section (A), T2w coronal section (B), T2w (C) and T1w (D) sagittal sections showing dilated suprasellar cistern compressing and displacing the pituitary gland inferiorly and the 3rd ventricle superiorly, which is secondarily dilated due to outflow obstruction.











The ADC map and DWI images show unrestricted diffusion in the region of dilated suprasellar cistern, suggesting the presence of an Arachnoid cyst.

Glucagon stimulation test

Time (min)	Cortisol (nmol/L)	GH
		(ng/ml)
0	355	0.24
60	271	0.15
90	566	0.46
120	726	2.9
150	747	1.3
180	666	0.45

Thyroid Function Test

FT4 - 19.6 pmol/L (10-28.2) TSH -1.68 mIU/L (0.46-4.62)

Conclusion



The case illustrates a possible association of suprasellar arachnoid cyst in BS not previously reported.



In children with Bartter syndrome who show persistent growth retardation despite optimizing medical therapy - is it growth hormone deficiency?







