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Synchronous symptomatic congenital choroid plexus lesions in siblings – To diagnose or to treat first?

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Background: We describe two siblings in a family born to consanguineous parents with bilateral choroid plexus lesions (CPL) for neurosurgical management. They presented to us synchronously in a few days interval with obstructive hydrocephalus. The aetiopathogenesis and management option with literature review is discussed.

Case Description: A 4-year-old girl presented with acute intraventricular haemorrhage from the CPL, whereas the 3-weeks-old male sibling presented with increasing drowsiness and sun-setting eyes in three days interval. The older sibling underwent an emergency ventriculostomy followed by selective choroidal artery embolization and then subsequently underwent a Ventriculoperitoneal (VP) shunt. The younger sibling was admitted after 3 days following his sister's presentation and underwent ventriculostomy and partial embolization of the left choroid. VP shunt was inserted after 2 weeks. He made good clinical recovery. Both were discharged home and are planned for diagnostic procedure for the future.

Discussion: The choroid plexus lesions account for 2-4% of paediatric brain tumours and have a neuroectodermal origin. Among the spectrum of lesions, the most common is choroid plexus papilloma and choroidal villous hypertrophy followed by carcinoma and atypical papiloma. They usually appear in the first two years of life.The aetiology of hydrocephalus is multifactorial. The treatment options include selective embolisation, endoscopic coagulation and surgical resection.The optimal managment in literature remains elusive. Genetic work up for parents and siblings is mandatory as a part of management to establish this correlation and also to predict similar occurrence for siblings in the future.

Conclusion: Congenital choroid plexus lesions in siblings are a rare occurrence and to our knowledge no similar cases have been described before. The primary management should be focussed on treating the secondary hydrocephalus and then to establish the diagnosis depending on the clinical picture. Follow up with CT and MRI is mandatory for monitoring and planning surgery.