Aqueductal stenosis in infancy

Aqueductal stenosis (AqS) is a frequent cause of congenital hydrocephalus (HCP) (up to 70% of cases [1]), but occasionally may be the result of HCP. Patients with congenital AqS usually have HCP at birth or develop it within ≈ 2–3 mos. Congenital AqS may be due to an X-linked recessive gene [2].

Four types of congenital AqS described by Russell (summarized [3]):

1. forking: multiple channels (often narrowed) with normal epithelial lining that do not meet, separated by normal nervous tissue. Usually associated with other congenital abnormalities (spina bifida, myelomeningocele)

2. periaqueductal gliosis: luminal narrowing due to subependymal astrocytic proliferation

3. true stenosis: aqueduct histologically normal

4. septum

1)
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2)
Matson DD. Neurosurgery of Infancy and Childhood. 2nd ed. Springfield: Charles C Thomas; 1969
3)
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