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Authors

Baram, Tallie Z
Fishman, Marvin A

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'Top of the Basilar' Artery Stroke in an Adolescent With Down's Syndrome

Tallie Z. Baram, MD, Marvin A. Fishman, MD

Basilar artery strokes are rare in children and adolescents.¹ DeVivo and Farrell, summarizing the world's literature up to 1972, found only five cases and added one of their own patients.² In all cases there appeared to be no predisposing causes or abnormalities of the posterior fossa vessels. The coexistence of Down's syndrome and vertebrobasilar strokes has not, to our knowledge, been reported. Down's syndrome is associated with cardiac malformations (mainly atrioventricular canal) that may lead to embolic strokes, and atlantoaxial dislocation that may also result in neurologic dysfunction. We have encountered a patient with trisomy 21 syndrome (without cardiac disease or atlantoaxial dislocation) who presented with classical clinical and computed tomographic evidence of a "top of the basilar" artery stroke.

REPORT OF A CASE

A 16-year-old youth with trisomy 21 syndrome was in his usual state of health when he suddenly staggered and collapsed, falling to the right. There was no involuntary movement, incontinence, or loss of consciousness. For two hours he "had no balance and complained of poor vision" and had violent nausea and vomiting. The episode completely resolved, only to recur in an almost identical fashion two weeks later; this time the deficits persisted.

On our examination, performed eight weeks after the second episode, the stigmata of Down's syndrome were present: broad flat facies; wide neck; low-set, posteriorly rotated ears; mongoloid-slanted palpebral fissures; and short hands with clinodactyly and simian creases. The rest of the physical examination, including the cardiovascular system, was unremarkable.

The patient's mental status examination revealed moderate mental retardation. The patient had visual agnosia, mainly for words, but no prosopagnosia. Other pertinent findings included an inability to sustain gaze to the left, bilateral end-gaze nystagmus that was both rotary and horizontal, a mild right central facial weakness, and poor palatal elevation. There was

slightly increased tone in the right upper and lower extremities and a mild right arm drift, with hyperreflexia and an extensor plantar response on the right. Striking dysmetria was noted bilaterally in both upper and lower extremities. The gait was very broad-based and ataxic.

A complete arteriogram performed one week after the second episode was normal. A computed tomographic scan confirmed bilateral cerebellar involvement, as well as both right and left occipital parietal lucencies (Figure). The findings were highly suggestive of infarction in the distribution of the superior cerebellar and posterior cerebral arteries.

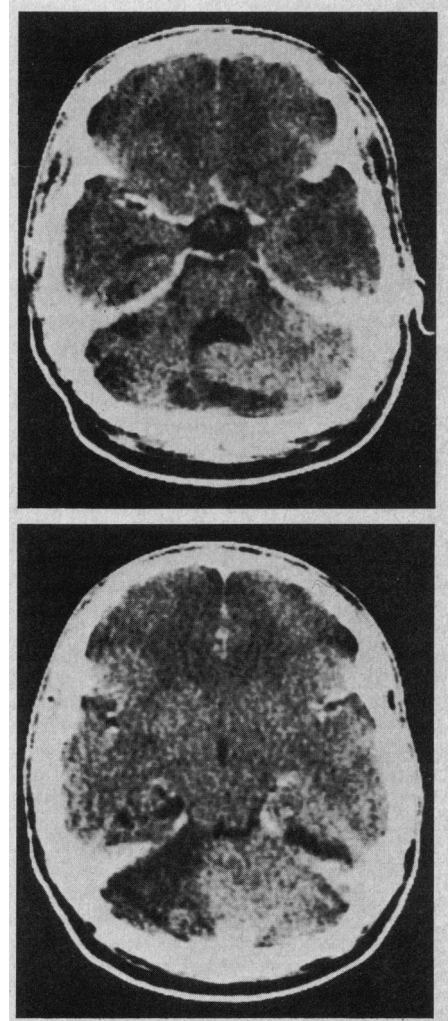
Results of a thorough cardiac evaluation, including clinical examination, electrocardiogram, echocardiogram, 24-hour rhythm monitor, and a treadmill stress test with rhythm monitoring were normal except for a mild mitral valve prolapse without hemodynamic significance. Results of the following additional studies were normal: electrolyte, total lipid, and cholesterol levels; the lipoprotein profile; thyroid functions; complete blood cell count (hematocrit, 50.6%); C3, C4, rheumatoid factor, and antinuclear antibody levels; and chest and spine roentgenograms, including complete and detailed flexion and extension views of the neck. Total triglyceride levels were moderately elevated, a frequent finding in Down's syndrome.³

COMMENT

Children and adolescents rarely present with posterior circulation strokes. When these do occur there is often a predisposing problem such as homocystinuria, moyamoya, cyanotic heart disease with paradoxical emboli, etc.

Down's syndrome may be associated with increased risk of strokes; the mechanism is cardiac emboli caused by right-to-left shunts or to valvular endocarditis. These emboli usually present as cerebral infarcts in the carotid distribution. Posterior circulation dysfunction, in patients with Down's syndrome, may result from kinking and occlusion of the vertebral arteries at the level of an atlantoaxial dislocation.

The patient manifested a top of the basilar artery syndrome without evidence of any spinal or cardiac abnormality. An arteriogram revealed no permanent occlusion of the vertebrobasilar system despite an obvious infarction visible on the computed tomographic scan. No congenital car-



Computed tomographic scan with contrast, demonstrating bilateral cerebellar and occipital lucencies.

diac lesion, arrhythmia, or endocarditis was found to account for his probable emboli stroke. Thus, the cause of this patient's stroke, as well as its relationship to his Down's syndrome, remain unclear at present. Further cases are needed before any causal relationship can be construed.

References

1. Swaimann KF, Wright FS: *Practice of Pediatric Neurology*. St Louis, CV Mosby Co, 1982.
2. DeVivo DC, Farrell FW Jr: Vertebrobasilar occlusive disease in children. *Arch Neurol* 1972; 26:278-281.
3. Salo MK, Solakivi-Jaakkula T, Kiviniemi T, et al: Plasma lipids and lipoproteins in Down's syndrome. *Scand J Clin Lab Invest* 1979;39:485-490.

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From the Departments of Neurology and Pediatrics, Baylor College of Medicine, Houston; and the Neurology Service, Texas Children's Hospital, Houston.

Reprint requests to Pediatric Neurology, Texas Children's Hospital, 6621 Fannin, Houston, TX 77030 (Dr Baram).