

- Halsey, J. H., Jr., Allen, N., Chamberlin, H. R. (1971) 'The morphogenesis of hydranencephaly.' *Journal of the Neurological Sciences*, **12**, 187-217.
- Holmes, L. B., Moser, H. W., Halldorsson, S., Mack, C., Pant, S. S., Matzilevich, B. (1972) *An Atlas of Diseases With Associated Physical Abnormalities*. New York: Macmillan.
- Marin-Padilla, M. (1970) 'Morphogenesis of anencephaly and related malformations.' *Current Topics in Pathology*, **51**, 145-174.
- McCormick, W. F. (1971) 'Is anencephaly a single entity? A brief note on the morphologic heterogeneity of anencephaly.' *Birth Defects*, **7**, 94-96.
- Minckler, J. (1971) *Pathology of the Nervous System*. New York: McGraw-Hill.
- Padget, D. H. (1972) 'Development of so-called dysraphism; with embryologic evidence of clinical Arnold-Chiari and Dandy-Walker malformations.' *Johns Hopkins Medical Journal*, **130**, 127-165.
- Pryor, H. B., Thelander, H. (1968) 'Abnormally small head size and intellect in children.' *Journal of Pediatrics*, **73**, 593-598.
- Smith, D. W. (1976) *Recognizable Patterns of Human Malformation*, 2nd Edn. Philadelphia: W. B. Saunders.
- Yakovlev, P. I. (1959) 'Pathoarchitectonic studies of cerebral malformations. III. Arrhinencephalies (holotelencephalies).' *Journal of Neuropathology and Experimental Neurology*, **18**, 22-55.

Bilateral Obstruction of the Vertebral Arteries in a Three-year-old Child

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Introduction

Occlusion of the vertebro-basilar arterial system is relatively rare in children. It occurs more frequently in adults due, in part, to the influence of arteriosclerosis and arthritic alterations in the vertebral column.

Fowler (1962) described necropsy findings from two cases of obstruction of the superior sections of the basilar artery in children. Dooley and Smith (1968) reported the angiographic findings of an obstructed basilar artery in a six-year-old child. A total of 18 children have been described in the literature, including the present case.

In some cases the occlusion occurs in the basilar artery or some of its principal branches and in others it occurs in the superior portion of the vertebral artery.

The primary reason for publishing this report is to present a child with complete obstruction of both vertebral arteries, a condition which, to our knowledge, has not been reported previously.

Case History

This three-year-old boy's gestation and birth had been normal. His tonsils had been removed at age two years. He had developed normally up to the age of three years when, without apparent cause, he developed a sudden speech difficulty and bilateral facial weakness, more predominant on the right. He had suffered no accidents or febrile illness to which the condition could be attributed. The neurological deficit lasted for 8 to 10 hours and was followed by complete recovery. Five days later he developed a 'jerking of the head', which was followed three to four hours afterwards by repetitive vomiting and gait disturbances, with instability and short steps. He also had profuse sweating and vertigo, and when laid down remained somnolent and asthenic. This condition lasted for three days, following which he was unable to talk and developed headache and bilateral horizontal nystagmus. He was then admitted to the Neuropaediatric Service.

On admission he was well oriented in time and space and answered all questions correctly, although he 'dragged' his syllables. He had generalized muscular hypotonia and showed active deep-tendon reflexes, especially in the lower limbs. He drooled slightly and showed bilateral facial paralysis. He was unable to stand up and tended to fall backwards: when maintained in the standing position by holding both arms, he stood with feet apart. Bilateral dysmetria and dysdiadochokinesia were observed, more severe on the right side. There was no visual disturbance and the other cranial nerves were normal.

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The most important laboratory data at that time were an erythrocyte sedimentation rate of 100 per hour and 8000 white cells. The heart examination and electrocardiogram were normal. An EEG showed diffuse slow-wave activity, without focal abnormalities.

On the fifth day of illness angiography of both carotid and vertebral arteries was performed. Carotid angiography showed coiling in the extra-cranial portion of both internal carotid arteries, a normal appearance of all intracranial branches of both internal carotid arteries, well-developed posterior communicating arteries and excellent retrograde filling of the superior portion of the basilar artery. Vertebral angiography demonstrated a complete obstruction of both vertebral arteries at the level of C2. Blood flow continued through several collateral branches which started on the right side from the same vertebral artery (Fig. 1) and on the left side from the vertebral and ascending cervical arteries (Fig. 2). There was good filling of the basilar artery and its principal branches along the entire course. The walls of the right vertebral artery had an irregular outline below the obstruction. The left vertebral artery appeared to be slightly hypoplastic below the obstruction, but regained its normal lumen upon receiving the collateral circulation. The treatment adopted was successful.

Evolution

After eight days the child was able to walk unassisted, although he was unstable. The facial weakness had disappeared. The erythrocyte sedimentation rate was 13 per hour.

One month later the child showed no clinical alterations. A year later the angiographic studies were repeated, in case there was recanalisation of the vertebral arteries: the arteries had a normal appearance. The child remains asymptomatic three years after the acute obstructive episode.

Discussion

Denny-Brown (1953) enumerated the symptoms of vertebro-basilar insufficiency in adults in the following order of frequency: (1) disturbance of equilibrium, (2) visual defects, (3) visceral disorders (*i.e.* vasomotor, cardio-respiratory), (4) severe headaches, predominantly occipital, (5) alterations of the state of consciousness, (6) motor difficulties, (7) auditory disorders, (8) facial or peripheral sensory disorders, (9) episodes very similar to

migraine, and (10) convulsions. The radiological alterations were well described by Schechter and Zingesser (1965).

The condition is infrequent in children and its aetiology is less specific and more variable than in adults, though the symptomatology is similar to that described by Denny-Brown (see Table). There is a clear predilection for males; of the 18 cases reported under 14 years of age only one has been female (Fowler 1962).

The obstruction was located in the basilar artery and in some of its branches in seven cases (Fowler 1962, Dooley and Smith 1968, Ouvrier and Hopkins 1970, Isler 1971, Moscow and Newton 1973); in the left vertebral and basilar arteries in three cases (Latchaw *et al.* 1974, Klein *et al.* 1976); in the right vertebral and basilar arteries in three cases (Harwood-Nash *et al.* 1971, Isler 1971, Singer *et al.* 1975); and in the two carotid arteries and the left vertebral artery in one case (Philippe *et al.* 1974). Two further cases presented a complete or almost complete obstruction of one vertebral artery and incomplete obstruction of the other vertebral artery (Ouvrier and Hopkins 1970, DeVivo and Farrell 1972). In one case the obstruction was not located (Richwein and Unger 1966). The present case is the first report of complete occlusion of both vertebral arteries.

It is difficult to suggest an aetiology since autopsies were performed in only three cases (Fowler 1962, Richwein and Unger 1966), in none of which had angiography been done. The occlusion was attributed to cranial trauma in two cases (Latchaw *et al.* 1974), to septicæmia with or without endocarditis in two cases (Fowler 1962, Ouvrier and Hopkins 1970), to congenital heart disease in one case (Richwein and Unger 1966), and in one case to alteration of the odontoid process with subluxation of C1–C2. In the majority of cases the aetiology was unknown, but



Fig. 1. Vertebral and carotid angiography on right side. Note coiling of extracranial internal carotid artery and complete obstruction of vertebral artery at level of C2. Collateral circulation starts from same vertebral artery.



Fig. 2. Vertebral angiography on left side. Note complete obstruction of hypoplastic vertebral artery at level of C2. Several collateral arteries have their origin in same vertebral artery and in ascending cervical artery.

the occlusion is usually attributed to an early disorder of arterial embryogenesis since the obstruction is verified at the level of C1 to C2 and there is important collateral circulation in the majority of cases. This collateral circulation derives from branches of the diseased vertebral artery which by-pass the site of obstruction, and also from branches of the ascending cervical artery and the occipital artery. In our patient the coiling of the two internal carotid arteries just above the bifurcation suggests arterial dysembryoplasia. However, the high erythrocyte sedimentation rate at the beginning of the disease also suggests inflammatory alterations of both dysplastic vertebral arteries.

Only three cases have resulted in death (Fowler 1962, Richwein and Unger 1966).

In five other children there were severe mental, motor and speech sequelae (Ouvrier and Hopkins 1970, Moscow and Newton 1973, Latchaw *et al.* 1974, Philippe *et al.* 1974, Singer *et al.* 1975). In a further six cases the sequelae were minimal and the children recovered almost completely (Dooley and Smith 1968, Ouvrier and Hopkins 1970, DeVivo and Farrell 1972, Latchaw *et al.* 1974, Klein *et al.* 1976). Complete recovery of functions was achieved by only two patients, Case 3 of Latchaw *et al.* (1974) and our own patient, though the former patient had a recurrence 15 years later at the age of 26, which was not severe. We are unaware of the outcome in one reported case.

We believe that the side of the arterial occlusion is important in the development

TABLE

<i>Author</i>	<i>Case</i>	<i>Sex</i>	<i>Age (yrs)</i>	<i>Clinical signs</i>	<i>Angiography</i>	<i>Outcome</i>
Fowler (1962)	1	F	1½	Relapsing pyramidal cranial nerve and speech disorders. Septicaemia. Coma five weeks after onset.	No	Death. Autopsy: thrombosis in superior portion of basilar artery.
	2	M	7½	Rapid onset of symptoms with pyramidal and cranial nerve disorders and coma.	No	Death. Autopsy: thrombosis in left superior communicating artery.
Richwein and Unger (1966)	1	M	13	Nausea, dizziness, pain on the left side of nose, deafness, right hemiparesis (Wallenberg's syndrome). Congenital heart lesion.	Embolism on vertebro-basilar circulation. No arteriographic or autopsy confirmation.	Death. Autopsy: thrombi in left heart.
Dooley and Smith (1970)	1	M	6	Left hemiparesis, vomiting, ataxia.	Occlusion of superior half of basilar artery.	Almost complete recovery.
Ouvrier and Hopkins (1970)	1	M	9	Vomiting, diplopia, vertigo, oculomotor nerve affection, dysarthria, right hemiparesis.	Occlusion of left vertebral artery. Narrowing of the lumen of right vertebral artery.	Moderate ataxia and dysarthria.
	2	M	4	Coughing, ataxia, dysarthria, right hemiparesis.	Occlusion of left vertebral artery.	Relatively insignificant motor sequelae.
Harwood-Nash <i>et al.</i> (1971)	3	M	14	Lethargy, tetraplegia, cardiopathy with bacterial endocarditis.	Occlusion of basilar artery.	Severe motor sequelae of cranial nerves and speech.
	1	?	?	?	Occlusion of right vertebral artery and of superior portion of basilar artery.	?
Isler (1971)	1	M	12	Dizziness, headache, nausea, vomiting, ataxia, hypalgnesia on the left side of face and tongue, diplopia, nystagmus (Wallenberg's syndrome).	Occlusion of basilar artery.	Almost complete recovery.
Devivo and Farrell (1972)	1	M	10	Visual defects, ataxia, dysarthria.	Occlusion of left vertebral artery and narrowing of lumen of right vertebral artery.	Almost complete recovery. (<i>continued</i>)

TABLE (cont.)

Author	Case	Sex	Age (yrs)	Clinical signs	Angiography	Outcome
Moscow and Newton (1973)	1	M	10	?	Occlusion of entire basilar artery.	Quadriplegia (5 years later).
Philippe <i>et al.</i> (1974)	1	M	17	Seizures, right hemiplegia.	Occlusion of left vertebral artery and of both carotid arteries.	Severe psychomotor alterations and seizures.
Latchaw <i>et al.</i> (1974)	1	M	7	Irritability, lethargy, visual disorders, right hemiparesis, decerebration.	Occlusion of basilar artery and narrowing of lumen of right vertebral artery.	Quadriplegia.
	2	M	11	Left hemiparesis, seizures, lethargy.	Occlusion of basilar artery and of left vertebral artery.	Discrete psychomotor sequelae.
	3	M	11	Headache, nausea, vomiting, lethargy, coma.	Occlusion of basilar artery and of left vertebral artery.	Total recuperation, but relapse occurred 15 years later.
Singer <i>et al.</i> (1975)	1	M	6	Headache, nausea, vomiting, ataxia, dysarthria, nystagmus, right hemiplegia.	Occlusion of right vertebral artery and of superior half of basilar artery.	Serious motor sequelae, and speech and visual sequelae.
Klein <i>et al.</i> (1976)	1	M	8	Vomiting, dizziness, staggering, difficulty in swallowing, headache, lethargy, nystagmus, hypalgnesia on left side of face, absent left corneal reflex, nasal voice, dysarthria, mild left hemiparesis, absent gag reflex bilaterally, left palatal weakness (Wallenberg's syndrome).	Occlusion of left vertebral artery. Hypoplastic left anterior inferior cerebellar artery.	Four months after the onset of syndrome, mild left facial hypalgnesia, nystagmus, left palatal weakness, occasional vertigo.
Pascual-Castroviejo <i>et al.</i> (present report)	1	M	3	Dysarthria, ataxia, profuse sweating, vomiting, fainting, paresis of the VIIth cranial nerve.	Complete occlusion of both vertebral arteries at level of C1-C2.	Total recovery.

CASE REPORTS

of the clinical picture. Obstruction of the basilar artery resulted in death in two cases (Fowler 1962) and severe neurological sequelae in Case 18 of Latchaw *et al.* (1974), Case 3 of Ouvrier and Hopkins (1970) and in the case reported by Singer *et al.* (1975). Only in Dooley and Smith's Case 3 was almost complete recovery achieved. Case 3 of Latchaw and colleagues, with basilar and left vertebral obstruction, cannot be appraised because angiography was performed at the relapse stage, 15 years after the first stroke.

A favourable outcome has occurred when the occlusion was located in one vertebral artery, as in the case reported by Klein *et al.* (1976) and in Ouvrier and

Hopkins' Case 2; or on both sides, as with DeVivo and Farrell's patient and Ouvrier and Hopkins' Case 1. Favourable outcome has also occurred when one of the vertebral arteries was partially permeable, and in our patient in whom both vertebral arteries were totally obstructed, but with adequate blood-flow through collateral branches.

AUTHORS' APPOINTMENTS

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SUMMARY

A three-year-old boy developed gait instability, nausea, vomiting, cranial nerve disturbances, hypotonus and dysarthria. Angiography of the four main cranial arteries showed complete obstruction of both vertebral arteries at the level of C1 to C2. Abundant collateral circulation was observed, which by-passed the obstruction to the vertebral arteries before their entrance into the posterior cranial fossa. The left vertebral artery was hypoplastic and both internal carotid arteries showed coiling in their extracranial portions. A high erythrocyte sedimentation rate at the beginning of the disease suggests an inflammatory alteration of both dysplastic vertebral arteries. The child recovered completely one month after the onset of symptoms.

All other reported cases of childhood vertebro-basilar obstruction are reviewed and it is emphasized that the site of arterial obstruction has an important bearing on the clinical outcome.

RÉSUMÉ

Obstruction bilatérale de l'artère vertébrale chez un enfant de trois ans

Les auteurs rapportent l'apparition chez un garçon de trois ans, d'une instabilité de la démarche, de nausées, de vomissements, de troubles des nerfs crâniens, d'hypotonie et de dysarthrie. Une artériographie des quatre principales artères crâniennes révèle une obstruction complète des deux artères vertébrales au niveau de C1-C2. Il fut observé une abondante circulation collatérale court-circuitant l'obstruction des artères vertébrales avant leur entrée dans la fosse crânienne postérieure. L'artère vertébrale gauche était hypoplasique. Les deux artères carotides internes présentaient un enroulement dans les portions intracrâniennes. L'enfant récupère complètement un mois après le début des troubles.

Il est fait une revue dans la littérature des obstructions vertébrobasilaires de l'enfance.

ZUSAMMENFASSUNG

Bilaterale Verlegung der A. vertebralis bei einem Kind im Alter von drei Jahren

Es wird über einen dreijährigen Jungen berichtet, der folgende Symptome entwickelte: Gangunsicherheit, Übelkeit, Erbrechen, Hirnnervenstörungen, Hypotonie und Dysarthrie. Die Angiographie der vier großen Hirngefäße zeigte einen kompletten Verschuß beider Aa. vertebrales in Höhe von C1-C2. Zahlreiche Kollateralkreisläufe sorgten für eine Umgehung der Obstruktion der Aa. vertebrales bis zu ihrem Eintritt in die hintere Schädelgrube. Die linke A. vertebralis war hypoplastisch. Die linke und rechte A. carotis interna waren in ihrem extracraniellen Anteil geschlängelt. Das Kind erholte sich vollständig einen Monat nach Einsetzen der Symptome.

Es wird ein Überblick der Literatur über die vertebro-basilar Obstruktion im Kindesalter gegeben.

RESUMEN

Presentamos un niño de tres años que desarrollaba inestabilidad en la marcha, náuseas, vómitos, alteraciones de los nervios craneales, hipotonía y disartria. La angiografía de las cuatro principales arterias cerebrales mostraba completa obstrucción de ambas arterias vertebrales al nivel de C1-C2. Se observaba abundante circulación colateral que comunicaba la zona pre con la postestenótica en ambas arterias vertebrales antes de su entrada en la fosa posterior. La arteria vertebral izquierda era hipoplásica. Ambas arterias carótidas internas presentaban coiling en su porción extracraneal. La velocidad de sedimentación elevada al comienzo del trastorno hace posible que hubiera alteraciones inflamatorias sobre ambas arterias vertebrales displásicas. El niño estaba completamente recuperado un mes después del comienzo de la sintomatología.

Se hace una revisión de los dieciocho casos de obstrucción vertebro-basilar y se hace incapié—en que la localización de la obstrucción es importante para la evolución clínica.

REFERENCES

- Denny-Brown, D. (1953) 'Basilar artery syndrome.' *Bulletin of the New England Medical Center*, **15**, 53-60.
- DeVivo, D. C., Farrell, F. W. (1972) 'Vertebrobasilar occlusive disease in children. A recognizable clinical entity.' *Archives of Neurology*, **26**, 278-281.
- Dooley, J. M., Smith, K. R. (1968) 'Occlusion of the basilar artery in a 6-year-old boy.' *Neurology*, **18**, 1034-1036.
- Fowler, M. (1962) 'Two cases of basilar artery occlusion in childhood.' *Archives of Disease in Childhood*, **37**, 78-81.
- Harwood-Nash, D. C., McDonald, P., Argent, W. (1971) 'Cerebral artery disease in children. An angiographic study of 40 cases.' *American Journal of Roentgenology*, **111**, 672-686.
- Isler, W. (1971) *Acute Hemiplegias and Hemisyndromes in Childhood. Clinics in Developmental Medicine*, No. 41/42. London: S.I.M.P. with Heinemann.
- Klein, R. A., Snyder, R. D., Sward, J. H. (1976) 'Lateral medullary syndrome in a child. Arteriographic confirmation of vertebral artery occlusion.' *Journal of the American Medical Association*, **235**, 940-941.
- Latchaw, R. E., Seeger, J. F., Gabrielsen, T. O. (1974) 'Vertebro-basilar arterial occlusion in children.' *Neuroradiology*, **8**, 141-147.
- Moscow, N. P., Newton, T. H. (1973) 'Angiographic implications in diagnosis and prognosis of basilar artery occlusion.' *American Journal of Roentgenology*, **119**, 597-604.
- Ouvrier, R. A., Hopkins, I. J. (1970) 'Occlusive diseases of the vertebrobasilar arterial system in childhood.' *Developmental Medicine and Child Neurology*, **12**, 186-192.
- Philippe, N., Floet, D., Rosenberg, D., Monnet, P. (1974) 'Obstruction des deux carotides internes et de la vertebrale gauche chez un nourrisson de 17 mois.' *Semaine des Hôpitaux de Paris, Annales de Pédiatrie*, **21**, 251-256.
- Richwein, R., Unger, G. (1966) 'Beobachtung einer Wallenberg-Syndrom im Kindesalter nebst Bemerkungen zum kindlichen cerebralen Gefäßprozess.' *Monatsschrift für Kinderheilkunde*, **8**, 442-443.
- Schechter, M. M., Zingesser, L. H. (1965) 'The radiology of basilar thrombosis.' *Radiology*, **85**, 23-32.
- Singer, W. D., Haller, J. S., Wolpert, S. M. (1975) 'Occlusive vertebrobasilar artery disease associated with cervical spine anomaly.' *American Journal of Diseases of Children*, **129**, 492-495.