Intracranial Dural Arteriovenous Shunts in Children

The Toronto Experience

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Introduction

Intracranial dural arteriovenous shunts (DAVS) are a group of different nosologic entities sharing a similar angioarchitecture¹. In the pediatric age group, three types of DAVS can be recognized (table 1)². The neonatal-type (NDAVS) is characterized by a malformation of the dural sinuses (figures 1A,B). The most common symptoms are related to increased intracranial pressure (ICP), mild cardiac failure and coagulation disorders. The reduction of venous outlets of the brain and additional overload into the sinuses lead to spontaneous thrombosis and consequent occlusion of all venous outlets as well as coagulation consumption syndromes.

The prognosis is poor for midline malformations whereas lateral malformations may have good outcomes if treated³.

The infantile-type (IDAVS), often present within the first few years of life, is often multifocal and the natural history includes a progressive occlusive venopathy with sigmoid sinus and/or jugular bulb stenosis and subsequent occlusion (figures 2A,B). Mild and well tolerated congestive heart failure (CHF) is usually present, and the treatment can be postponed until the development of macrocrania or hydrodynamic disorders. At that point, treatment is mandatory to avoid mental retardation.

Other neurological symptoms will also depend on whether the cavernous sinus has matured sufficiently to be able to capture the cortical venous system of the brain (natural by-pass to the venous outflow)¹⁻³.

The adult-type (ADAVS) is similar in appearance and presentation to DAVS in the adult population. It can develop following thrombosis of the sinus wall or after traumatic injuries to the sinus. Its symptoms depend on the drainage patterns of the shunt and on the coexistence of venous thrombosis. Multifocal and extra-sinusal (duro-subdural, osteodural) ADAVS have not been described in children^{2,3}. We report a series of 11 cases with DAVS, describe their treatment and report their outcomes.

Methods

From April 1986 to July 2003, 11 children (nine boys and two girls) were evaluated for DAVS in our institution (table 2). These patients ranged in age from 5 days to 12 years at the time of first evaluation.

A retrospective review of the patient's charts and of all available imaging studies was performed. Their follow up was obtained in clinic consultations.

The treatment options were either transarterial embolisation alone, transvenous embolisa-

Figure 1 Neonatal-type DAVS (Case 5) A) Right occipital arteriogram (AP view) shows fistula from the occipital artery into a large, malformed dural sinus. B) Venous phase of right occipital arteriogram (lateral view) shows an occluded right transverse sinus (arrowhead) and a stenotic left jugular bulb (arrowhead).

tion alone or both routes combined. N-butylcyanoacrylate (NBCA) and Polyvinyl Alcohol (PVA) were the agents of choice in transarterial approaches, whereas coils were preferred in transvenous embolisations.

The embolisation was classified as partial when there was evidence of a residual arteriovenous shunt on angiography.

Results

All patients treated ($n = 10$) had embolisation only. One patient (Case 9), with a Factor VIIA deficiency, was not treated. There were 2 NADVS, 4 IDAVS and 5 ADAVS. The most common initial clinical presentations included bruit ($n = 5$), headaches ($n = 5$), neurological deficits ($n = 3$), CHF ($n = 3$), seizures ($n = 2$),

Table 1 Classification of DAVS in the pediatric population^{2*}

Type	Name	Description
1	Neonatal DAVS	Giant dural lake with a slow communication towards the other sinuses
2	Infantile DAVS	Often multifocal, without sinus malformation, with high-flow angiopathy on both the arterial and the venous side (resulting in jugular bulb occlusion)
3	Adult DAVS	Cavernous plexus or sigmoid sinus, sinuses may be partially thrombosed

*Modified from Lasjaunias, P.L. and K. ter Brugge, *Vascular Diseases in Neonates, Infants and Children. Interventional Neuroradiology Management*, ed. P.L. Lasjaunias. 1997, Paris: Springer-Verlag Berlin Heidelberg

Table 2 Clinical and radiological findings in 11 children presenting with DAVS

Case (No.)/Sex	Age at First Treatment	Presentation	Type	Other Clinical Information	Initial Brain Imaging	DAVS Location
1/ M	2 years	Cranial bruit (right retromastoid region)	3	...	MRI: normal	Right sigmoid sinus
2/ M	5 years	Right hemiparesis (TIA*), headaches, seizures	2	...	CT: large right frontal DVA†, intracerebral haematoma	Multifocal SSS‡
3/ M	4 years	CHF§, headaches, seizures, left-sided neck and cranial bruit, macrocrania	2	Severe asthma	CT: moderate hydrocephalus	Multifocal left superior petrosal sinus
4/ F	12 years	Bruit in the left ear, headaches, occasional right hand numbness	3	...	MRI: normal	Left transverse sinus
5/ M	1 year	Headaches, epistaxis, macrocrania	1	Prominent orbital veins	MRI: moderate hydrocephalus, dystrophic brain calcifications	Torcular
6/ M	6 months	Developmental delay and left-sided facial swelling, multiple left ear haemorrhages	2	Heart murmur caused by a patent ductus arteriosus	MRI: mild brain atrophy	Multifocal left sphenoparietal and inferior petrosal sinuses
7/ M	5 days	CHF	1	...	MRI: mild hydrocephalus	SSS
8/ M	2 months	CHF and left-sided cranial bruit	2	on digoxine	MRI: normal	Multifocal left transverse sinus
9/ M	7 years	Right proptosis and chemosis	3	Factor VIIA deficiency, mild head trauma 6 years before	MRI: normal, prominent right ophthalmic vein	Right cavernous and inferior petrosal sinuses
10/ M	10 years	Bilateral proptosis (Right>Left) and chemosis	3	Down's syndrome	...	Right ophthalmic vein
11/ F	3 years	Cranial bruit and headache	3	Right cheek and neck haemangioma	MRI: normal	Right sigmoid sinus

*TIA, Transient Ischaemic Attack; †DVA, Developmental Venous Anomaly;
‡SSS, Superior Sagittal Sinus; §CHF, Congestive Heart Failure.

proptosis ($n = 2$), and bleeding ($n = 2$). One patient with an adult-type DAVS (Case 9), had a history of trauma (table 3). CHF was detected immediately after birth in all three cases. The 2 cases of severe CHF occurred in the IDAVS. The third patient had mild CHF and a NADVS.

On the initial brain imaging, normal brain parenchyma was present in 6 patients, whereas hydrocephalus was present in 3 cases (two cases of NADVS and one case of IDAVS). Multifocal DAVS were found in all cases of IDAVS. Three of the 4 multifocal DAVS involved either the transverse or the petrosal sinuses, and one involved the superior sagittal sinus. The sigmoid sinus was involved in 2 of the 5 cases of ADAVS.

A total of 24 sessions of embolisation were done in 10 patients. The number of sessions per patient ranged from 1 to 5 (mean = 2.4 per patient). All patients were treated using transarterial embolisation (NBCA, PVA or Gelfoam). Four patients were also treated using transvenous embolisation (coils, stent or balloon). No patient was treated using a direct puncture of the involved sinus.

Complete obliteration of the DAVS was achieved in 3 ADAVS. Two of them were treated in a single session.

There were no complications from the endovascular treatment, although developmental delay became evident after partial embolisation in one IDAVS (Case 2) and in one

Table 3 Treatment and Outcome in 10 children with DAVS*

Case #	Embol†. Route	# of Embol. Sessions	Embol. Materials	Angiographic Outcome (at last Embol.)	Clinical Outcome (time from 1st Embol.)
1	Combined‡	2	NBCA§, Coils	Cure	Cured, no developmental delay (7 years)
2	Combined	4	NBCA, Coils, Stent, Balloon	Partial., recurrent left internal jugular stenosis	Controlled seizures, mild developmental delay, progressive cortical atrophy and dystrophic calcifications (3 years)
3	Combined	2	NBCA, Coils	Partial	Death (1 month)
4	Transarterial	5	NBCA, PVA — —, Gelfoam	Partial	Persistent symptoms (8 years)
5	Transarterial	1	NBCA, PVA	Partial	Persistent symptoms, developmental delay (5 years)
6	Transarterial	4	NBCA, PVA	Partial	Relieved symptoms, no developmental delay (4 years)
7	Transarterial	1	NBCA	Partial	Persistent symptoms (1 year)
8	Combined	3	NBCA, Coils	Partial	Persistent symptoms (17 years)
10	Transarterial	1	NBCA	Cure	Cured (5 years)
11	Transarterial	1	NBCA	Cure	Cured (4 years)

*Case 9 excluded, not treated; †Embol., Embolization; ‡Combined, Transarterial and Transvenous embolization; §NBCA, N-butylcyanoacrylate; — — PVA, Polyvinyl Alcohol

Figure 2 Infantile-type DAVS (Case 2). A) Left internal carotid arteriogram (lateral view) shows a multifocal DAVS (arrows) towards the superior sagittal sinus. B) Left external carotid arteriogram (lateral view) shows separate fistula into the superior sagittal sinus and transverse sinus (arrows).

NDAVS (Case 5). One IDAVS (Case 3) died, 15 days after being discharged, due to respiratory complications and seizures. In 5 patients symptoms persisted after partial embolisation (follow-up from one to 17 years), although headaches, seizures and cardiac function were well controlled in all cases. Even though Case 6 had an excellent clinical and neurological recovery at 4 years, a longer follow-up is necessary to classify it as a cure, since the IDVAS was partially embolised.

Discussion

DAVS account for approximately 10% of all intracranial arteriovenous shunts in children⁴, are often multifocal and have a more aggressive clinical course compared to adults^{5,6}. The NDAVS has a poor outcome due to the development of venous outlet obstruction and coagulation consumption syndromes^{2,3}. In our two NDAVS, both affecting the superior sagittal sinus and the torcula, the outcome was not favourable, with persistent symptoms in both and developmental delay in one.

Four of our 11 patients had multifocal lesions, typical of the IDAVS. Two of the four had a poor outcome despite treatment (developmental delay in one and death in another). In the patient with persistent developmental delay there was mild brain atrophy at the time of diagnosis.

Two of these 4 patients with IDAVS had

seizures, two had hydrocephalus and one had macrocrania. One of the patients who had developmental delay had angioplasty and stenting of a jugular bulb stenosis. Initially this improved his neurological signs and symptoms however the stenosis recurred and his neurological function deteriorated.

Repeat angioplasty resulted in only a temporary improvement in the venous congestive encephalopathy.

The ADAVS have the best prognosis. These often do not have brain symptoms at presentation⁷⁻⁹. In our five cases of ADAVS, the presentations included bruit in three and orbital congestion in two. Transarterial embolisation was curative in three of the four patients in this group.

Conclusions

The current treatment of choice for DAVS in children in endovascular therapy, which may be life saving in the setting of cardiac failure and curative in ADAVS. Treatment strategies should focus on the patient's clinical status, and on the neurological prognosis.

The goal of treatment may be to arrest the neurological deterioration since angiographic cure may not be feasible. Venous approaches have to be considered with caution, since progressive venous occlusive phenomena may interfere with the drainage pattern of the healthy brain.

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