SVN Stroke & Vascular Neurology

Paediatric intracranial dural arteriovenous fistulas: clinical characteristics, treatment outcomes and prognosis

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To cite: Su X, Ma Y, Song Z, *et al*. Paediatric intracranial dural arteriovenous fistulas: clinical characteristics, treatment outcomes and prognosis. *Stroke & Vascular Neurology* 2025;**10**: e003122. doi:10.1136/svn-2024-003122

► Additional supplemental material is published online only. To view, please visit the journal online (https://doi.org/10.1136/svn-2024-003122).

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Received 13 January 2024
Accepted 9 May 2024
Published Online First 6 June 2024

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ABSTRACT

Background Compared with dural arteriovenous fistulas (DAVFs) in adult, paediatric DAVFs are notable for distinct clinical manifestations, low cure rate and poor prognosis. However, due to the limitations of small sample sizes, the long-term prognosis and follow-up data have not been described.

Methods Clinical data from 43 consecutive paediatric DAVFs were documented and analysed between 2002 and 2022 at the author's institution. They were divided into infantile (Lasjaunias classification) and non-infantile (adult type and dural sinus malformation (DSM)) type DAVFs based on prognosis differences.

Results Their mean age at first symptoms was 8.4±6.0 years. 29 boys and 14 girls presented between at birth and 18 years of age. 5 of 10 patients ≤1 year of age presented with asymptomatic cardiomegaly compared with 5/33 patients >1 year of age (p=0.022). 42 (88.4%) patients received endovascular treatment alone, while 9.3% underwent radiosurgery, burr hole embolisation or surgery. 28 (65.1%) patients experienced DAVF obliteration by the end of treatment. Among them, 26 cases underwent embolisation alone, one case had embolisation in conjunction with surgery, and one case underwent burr hole embolisation. The overall complication rate among patients was 9.3%, all resulting from endovascular treatment. According to the Lasjaunias Classification, there were 18 cases of adult type, 17 cases of infantile type and 8 cases of DSM. Compared with non-infantile-type DAVFs, infantile-type DAVFs showed more times of treatment, lower cure rate and worse prognosis (p<0.001, 0.003 and 0.021, respectively). The average follow-up duration was 41.4±36.2 months (3–228 months). 8 (22.9%) patients died.

Conclusions Most adult-type DAVFs and DSMs can now be effectively treated with embolisation, resulting in good outcomes and prognosis. However, there are still challenges in treating infantile-type DAVFs, and the prognosis is frequently poor.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Paediatric dural arteriovenous fistulas (DAVFs) are notable for their rarity, distinct clinical manifestations (congestive heart failure, facial vein prominence, hydrocephalus, macrocrania, developmental delay), low cure rate and poor prognosis in some case series or reports.

⇒ The usual descriptions and adult classifications of DAVFs (Cognard and Borden classification) are not practically applicable. Lasjaunias classified them according to angioarchitecture and natural course: infantile type/multifocal DAVFs, adult-type DAVFs and dural sinus malformations (DSMs).

⇒ The success rate of treatment and clinical outcomes of infantile-type DAVFs are not satisfactory.

WHAT THIS STUDY ADDS

⇒ This is the largest single-center study on the topic of paediatric DAVFs with detailed information.

⇒ Adult-type DAVFs and DMSs can be treated successfully with current techniques, but treating infantile-type DAVFs remains difficult.

⇒ The long-term follow-up results indicate that infantile-type DAVFs has a poorer prognosis. It is extremely necessary to closely monitor them in the long term.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ Most adult-type DAVFs and DSMs can now be effectively treated with embolisation, resulting in good outcomes and prognosis. However, there are still challenges in treating infantile-type DAVFs, and the prognosis is frequently poor. Improved comprehension regarding their pathogenesis, combined with targeted antiangiogenic treatments, may have the potential to enhance the outcome of these patients.

INTRODUCTION

Intracranial dural arteriovenous fistulas (DAVFs) are abnormal shunts between the dural arteries or pial arteries and dural venous sinus or cortical veins. They account for approximately 10%–15% of all intracranial vascular malformations.[1 2] Paediatric DAVFs are notable for their rarity, distinct clinical manifestations, low cure rate and poor prognosis.[3–12] It accounts for approximately 10%–14% of all intracranial arteriovenous shunts in children.[6 13] Distinct clinical manifestations mainly include congestive heart

failure, facial vein prominence, hydrocephalus, macrocrania and developmental delay.[3–5 7 8] A 1996 study of 29 paediatric patients with DAVFs found a 31% mortality rate, a favourable outcome in 45% of patients and an unfavourable outcome in 24% of patients.[8] A recent systematic literature review of paediatric intracranial DAVFs including 61 studies with 69 individual patients showed that only half of DAVFs were completely obliterated.[3] Compared with adult DAVFs, paediatric DAVFs are more fatal.[3 14] Understanding paediatric DAVFs comprehensively is crucial, but most of the prior research has been based on case reports or small sample studies. Here, we present 43 cases of paediatric DAVFs that were treated at our centre over a span of 20 years, aiming to raise awareness and understanding of this DAVF subtype.

METHODS

Study design

This was a retrospective study of 43 patients with paediatric DAVFs (diagnosed as DAVFs at <19 years of age) who were admitted to one hospital between 2002 and 2022. The ethics committee of our hospital approved this study. The study followed the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE guidelines).[15]

Data collection and definition

Patients' medical records, imaging reports and angiographic data were maintained (table 1 and online supplemental table 1). Age, gender, clinical presentation, angiographic findings, treatment modalities, angiographic and clinical outcomes and complications were reviewed. Admission, outpatient visits and telephone follow-up were all considered. Most patients were contacted by telephone to check on their status after treatment. Cardiomegaly was determined through a chest X-ray report. Radiological follow-up included magnetic resonance angiography (MRA) and digital subtraction angiography (DSA). Persistent fetal venous structures included falcine sinus and occipital sinus. Complete occlusion was defined as the absence of visualisation of the sinus or shunted cortical vein in the delayed venous phase on the completion angiogram. Subtotal occlusion was defined as more than 90% of the lesions disappeared but not completely occluded. Partial occlusion was defined as less than 90% resolution of the lesion after treatment. We categorised paediatric DAVFs based on the follow classifications: (1) Cognard classification,[1] (2) Borden classification[2] and (3) Lasjaunias classification: infantile type/multifocal DAVFs, adult type and dural sinus malformation (DSM).[8]

Angiographic images were reviewed by three neurosurgeons (XS, YM and ZS). Three experienced neurosurgeons (M, HZ and PZ) independently verified the angiographic characteristics, results of embolisation and MRA and DSA follow-up imaging. If there were any disagreements, a discussion would be held in order to reach an agreement.

Table 1 A brief summary of the angioarchitecture of the paediatric dural arteriovenous fistula

Variable	Value
No. of patients	43
Male	29 (67.3%)
Age first symptoms (years, mean±SD)	8.4 (6.0)
Cognard type	
Cognard type I	6 (14.0%)
Cognard type IIa	5 (11.6%)
Cognard type IIb	9 (20.9%)
Cognard type IIa+b	15 (34.9%)
Cognard type III	4 (9.3%)
Cognard type IV	3 (7.0%)
Cognard type V	1 (2.3%)
Borden type	
Borden type I	11 (25.6%)
Borden type II	24 (55.8%)
Borden type III	8 (18.6%)
Lasjaunias type	
Infantile type	17 (39.5%)
Adult type	18 (41.9%)
Dural sinus malformation	8 (18.6%)

Treatment and outcomes

Patients with paediatric DAVFs were managed through different approaches, including conservative treatment, endovascular intervention, a combination of endovascular and surgical intervention or a combination of endovascular and gamma knife radiosurgery (table 2).

Two investigators (XS and YM) reviewed medical records from each patient's most recent clinical follow-up evaluation or phone consultation to determine neurological status and developmental disability; any disagreements were resolved through additional review and consensus. All patients were evaluated using the modified Rankin Scale (mRS) score during the preoperative, postoperative and follow-up periods. A good clinical outcome was defined as a mRS score of 0–2. The duration of follow-up was calculated from the last treatment to the final clinical, imaging report, or telephone consultations.

Statistical analysis

Our cohort of patients was stratified by Lasjaunias classification due to the treatment difficulties and poor prognosis of infantile DAVFs[4 7 8]: infantile type and non-infantile type (adult type and DSM) (table 3 and figure 1). Descriptive statistics were calculated using Excel 2016 (Microsoft). SPSS for Windows (V.26.0; International Business Machines Corporation) and GraphPad Prism V.9.0 was used for statistical analysis. Variables are expressed as the mean±SD, the median (P25, P75) or the number of patients (percentage). The Kaplan-Meier

Table 2 Treatment and results of paediatric dural arteriovenous fistulas

Variable	Value
No. of patients	43
Untreated	1 (2.3%)
Treated	42 (97.7%)
Embolisation alone	38 (88.4%)
Burr hole embolisation	1 (2.3%)
Embolisation+surgery	2 (4.7%)
Embolisation+radiosurgery	1 (2.3%)
Median no. of treatment (range)	1 (0–7)
Complete embolisation	28/42 (66.7%)
Subtotal embolisation	7/42 (16.7%)
Partial embolisation	7/42 (16.7%)
Follow-up duration*	
Mean±SD	41.4±36.2 months
Median	16 months
Range	3–228 months
Median last mRS score (range)†	1 (0–6)
Good outcomes (last mRS score 0–2)†	24/35 (68.6%)
Death†	8/35 (22.9)
Complications	4 (9.3)

*Data available for 34/43 patients.
†Data available for 35/43 patients.
mRS, modified Rankin Scale.

method was used to depict the probability of survival rate, and the curves were compared using the log-rank test. Pearson's χ^2 test (applying correction by continuity if necessary) or Fisher's exact test was used for categorical variables and Wilcoxon rank-sum (Mann-Whitney) tests were used to compare ordinal data to evaluate differences in clinical variables and outcomes. Statistical significance was assigned for $p<0.05$.

RESULTS

Patient baseline characteristics

Of the 43 patients included for analysis, 10 (23.3%) presented the first related symptoms at ≤1 year of age and 33 presented at >1 year of age (median age 8.5 years, range at birth to 18.7 years). Five (11.6%) were diagnosed as DAVFs at ≤1 year of age and 38 were diagnosed at >1 year of age (median age 10.6 years, range at 1.0 years–18.9 years). Their mean age at first symptoms was 8.4±6.0 years. Twenty-nine (67.4%) boys presented at a median age of 9.6 years and 14 girls presented at a median age of 4 years. Ocular symptoms (proptosis, chemosis, diplopia, visual deterioration and retro-orbital pain) (17/43, 39.5%), headache (10/43, 23.3%), pulsatile head mass/dilated facial veins (9/43, 20.9%), focal neurological deficits (haemiparesis, aphasia and facial droop) (7/43, 16.3%), seizures (7/43, 16.3%), developmental delay (5/43, 11.6%), tinnitus (4/43, 9.3%), and dizziness (4/43, 9.3%) were the main symptoms.

Congestive heart failure was not observed in any of the 10 (23.3%) patients who had cardiomegaly, but one had congenital heart disease (ventricular septal defect). Among these cases, five (50%) were ≤1 year of age, six (60%) belonged to the infantile type group and two belonged to the DSM group. Five (11.6%) patients had hydrocephalus or macrocephaly. Three (60%) of them had at ≤1 year of age, two (40%) belonged to the infantile type group and two (40%) belonged to the DSM group. Seven (16.3%) patients presented with neurological deficits. Among them, two (28.6%) presented at ≤1 year of age, four (57.1%) belonged to the infantile type group and three belonged to the DSM group. Seven (16.3%) had intracranial haemorrhage and none of them presented at ≤1 year of age. Among them, three (42.9%) belonged to the infantile type group and three belonged to the adult type group. Five (11.6%) had developmental delay and three (60%) had at ≤1 year of age. Among them, three (60%) belonged to the infantile type group and two belonged to the DSM group. Comorbidities were present in 11 (25.6%) of the patients, including scalp, face and intracranial arteriovenous malformations (five patients), lymphatic malformation (one patient), head haemangiomas (two patients), venous malformation (one patient), cerebellar tonsillar herniation and cerebral cavernous malformation (one patient) and neurofibromatosis (one patient). Among them, three (27.3%) had at ≤1 year of age, five (45.5%) belonged to the infantile type group and five belonged to the adult type group. Two (4.7%) patients had history of head trauma. Both of them presented at >1 year of age and belonged to adult type group. Significant differences in cardiomegaly were observed between the patients ≤1 year old versus patients >1 year old (5/10, 50% vs 5/33, 15.2%; p=0.022) but not in other presentations (not shown in table). Significant differences in pulsatile head mass were observed between the infantile type versus non-infantile type (p=0.024) but not in other presentations (table 3). The baseline characteristics of the 43 patients are summarised in table 1 and online supplemental table 1.

DAVF angioarchitecture

Multiple locations of DAVFs in a patient occurred in 30.3% (10/33) of the patients >1 year old versus 10% (1/10) of patients ≤1 year old (p=0.381). Common DAVF locations included the transverse-sigmoid sinus (n=18), convexity/superior sagittal sinus (n=11), torcular (n=7), cavernous sinus (n=5), tentorial (n=4), middle cranial fossa (n=2), anterior cranial fossa (n=1) and other location (n=10) (occipital sinus, intraorbital, jugular foramen/hypoglossal canal). Three of them were isolated sinus DAVFs. Some specific locations of paediatric DAVFs have been reported in previously published articles.[16–18] Both age groups had similar percentages of patients with venous ectasia, venous reflux, venous sinus thrombus/occlusion

Table 3 Population, fistula angioarchitecture and treatments of paediatric DAVF stratified by Lasjaunias type

Variables	Total (n=43)	Infantile type (n=17)	Non-infantile type (n=26)	P value*
Age at first symptoms	8.4±6.0	8.9±5.6	8.1±6.2	
Males (%)	29 (67.4%)	10 (58.8%)	19 (73.1%)	0.329
Headache	10 (23.3%)	5 (29.4%)	5 (19.2%)	0.440
Ocular symptoms	17 (39.5%)	4 (23.5%)	13 (50%)	0.157
Pulsatile head mass	9 (20.9%)	7 (41.2%)	2 (7.7%)	**0.024¶**
Seizures	7 (16.3%)	3 (17.6%)	4 (15.4%)	1.000
Focal neurological deficit	7 (16.3%)	4 (23.5%)	3 (11.5%)	0.536
Haemorrhage	7 (16.3%)	3 (17.6%)	4 (15.4%)	1.000
Cardiomegaly	10 (23.3%)	6 (35.3%)	4 (15.4%)	0.254
Developmental delay	5 (11.6%)	3 (17.6%)	2 (7.7%)	0.611
Hydrocephalus	4 (9.3%)	1 (5.9%)	3 (11.5%)	0.930
Trauma	2 (4.7%)	0	2 (7.7%)	0.511†
High grade DAVFs	32 (74.4%)	15 (88.2%)	17 (65.4%)	0.186
Comorbidity	12 (27.9%)	5 (29.4%)	7 (26.9%)	0.859
Multifocal lesions	11 (25.6%)	10 (58.8%)	1 (3.8%)	**<0.001¶**
Venous ectasia\varix	28 (65.1%)	11 (64.7%)	17 (65.4%)	0.964
Venous reflux	32 (74.4%)	15 (88.2%)	17 (65.4%)	0.186
Venous sinus thrombus/occlusion	26 (60.5%)	10 (58.8%)	16 (61.5%)	0.859
Venous sinus dilation	19 (44.2%)	8 (47.1%)	11 (42.3%)	0.759
Persistent fetal venous structures	8 (18.6%)	5 (29.4%)	3 (11.5%)	0.284
Median no. of treatment (range)	1 (0–7)	3 (1–7)	1 (0–3)	**<0.001‡¶**
Complications	4 (9.3%)	2 (11.8%)	2 (7.7%)	1.000
Complete occlusion	28 (65.1%)	6 (35.3%)	22 (84.6%)	**0.003¶**
Death§	8/35 (22.9%)	6/14 (42.9%)	2/21 (9.5%)	0.059
Median last mRS score (range)§	1 (0–6)	3.5 (0–6)	1 (0–6)	**0.004‡¶**
Good outcomes (last mRS score 0–2)§	24/35 (68.6%)	6/14 (42.9%)	18/21 (85.7%)	**0.021¶**

*P values are derived from Pearson's χ^2 test (applying correction by continuity if necessary) unless otherwise specified.
†Fisher's exact test.
‡Wilcoxon log rank-sum (Mann-Whitney) test.
§Data available for 35/43 patients.
¶Bold values represent statistical significance.
DAVFs, dural arteriovenous fistulas; mRS, modified Rankin Scale.

and venous sinus dilation (not shown in table). There were no significant differences in venous ectasia, venous reflux, venous sinus thrombus/occlusion and venous sinus dilation between infantile type and non-infantile type (table 3). Detailed informations about the angioarchitecture can be found in table 1, online supplemental table 1 and table 3.

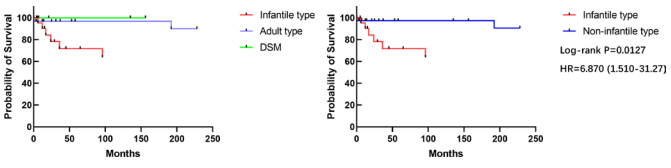

Figure 1 Kaplan-Meier curves demonstrating probability of survival rates for all patients as the follow-up time in months. Kaplan-Meier curves (log-rank test) illustrating probability of survival rates of paediatric DAVFs in non-infantile-type DAVFs were higher than infantile-type DAVFs. DAVFs, dural arteriovenous fistulas; DSM, dural sinus malformation.

Treatment and outcomes

Forty-two patients received between 1 and 7 endovascular, radiosurgical or surgery procedures with a median of two interventions. A paediatric patient received conservative treatment after multiple failed attempts at femoral artery puncture resulted in femoral artery dissection. About 88.4% of our patients received endovascular treatment only. In terms of the number of treatments, no significant difference was found between different age groups (not shown in table). However, a significant difference was found between the infantile type and non-infantile type (p<0.001; table 3). All patients who were treated received

embolisation treatment. The treatment material mainly included: coils, 13/42 patients (40%); Glubran/nBCA, 22/42 patients (52.4%); Onyx, 36/42 patients (85.7%); balloons, 14/42 patients (33.3%). Two (4.7%) patients underwent craniotomy and resection following embolisation. One (2.3%) patient underwent embolisation following burr hole access for transverse sinus and one patient underwent stereotactic radiosurgery following embolisation.

Complete embolisation was achieved in 28 (66.7%) of 42 patients who received treatment. Among them, 26 cases underwent embolisation alone (92.8%), 1 case had embolisation in conjunction with craniotomy (3.6%) and 1 case underwent burr hole embolisation (3.6%). Unfavourable clinical outcomes occurred in 57.1% (8/14) of infantile-type DAVFs, 16.7% (1/6) of DSM with DAVFs and 13.3% (2/15) of adult-type DAVFs. The complete occlusion rate was 35.3% (6/17), 75% (6/8), 88.9% (16/18) in infantile, DSM and adult-type DAVFs, respectively. There was no significant difference between the patients ≤1 year old and patients >1 year old (7/10, 70% vs 21/33, 63.6%). However, a significant difference was found between the infantile type and non-infantile type in complete occlusion rate (p=0.003; table 3).

Out of the 43 patients, 4 (9.3%) experienced procedural complications, all resulting from endovascular treatment. These complications occurred in patients who were in 2.0 years, 9.0 years, 9.5 years and 9.9 years old. Complications observed during the procedure involved femoral artery dissection and arterial infarction. The underlying cause was identified as the penetration of embolic agents across external–internal anastomoses into the cerebral parenchymal circulation. There was no significant difference in complication rate between patients ≤1 year old and patients >1 year old, infantile type and non-infantile type.

Follow-up

The average follow-up duration was 41.4±36.2 months, with a median of 16 months and a range of 3 months to 228 months. Twenty (46.5%) of 43 patients demonstrated imaging elimination of DAVF at last imaging follow-up (16 with DSA and 4 with MRA).

After the last treatment, 8 (18.6%) patients were lost to follow-up. The median mRS score at last follow-up for patients (35/43) was 1. Good clinical outcome (mRS score of 0–2 at last clinical follow-up) was documented in 6/8 patients ≤1 year old at presentation as compared with 18/27 patients >1 year old (p=1.000). However, a significant difference was found between the infantile type and non-infantile type in good clinical outcomes (p=0.021; table 3). At last follow-up, 8/35 (22.9%) patients died. One patient died within 1 week of treatment due to an arterial infarction, and one died 1 year later due to multiple organ failure. Four patients died as a result of intracranial haemorrhage after partial and subtotal embolisation. Two patients died for unknown reasons 3 and 16 years after the complete embolisation. There was no significant difference in death rates between patients ≤1 year old and patients >1 year old, but the infantile type group had a higher death rates than the non-infantile type group, though this difference was not significant (p=0.059; table 3). The Kaplan-Meier curves (log-rank test) illustrating the probabilities of survival of paediatric DAVFs in the non-infantile type group were higher than in the infantile type group (figure 1). Detailed informations about the treatment and follow-up can be found in tables 2 and 3.

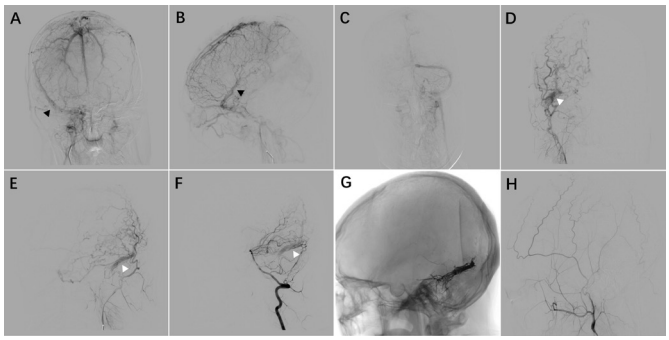

Figure 2 A paediatric patient in age range of 10 presented with headache, bilateral impaired vision and papilledema. (A–C) The sigmoid sinus and transverse sinus were occluded (venous sinus thrombosis) on the right side and the superficial cerebral vein (A and B, black arrow heads) was mainly involved in venous drainage. On the left side, the sigmoid sinus and transverse sinus were patent (C). Note the marked venous congestion (B). (D–F) Right external carotid artery and vertebral artery angiograms revealed a right Cognard III/Borden III/adult type isolated transverse sinus (E and F, white arrow heads) dural arteriovenous fistula mainly supplied by right posterior branch and petrosal branch of the middle meningeal artery, right transosseous branches of the left occipital artery, the right posterior meningeal artery and posterior cerebral artery with cortical veins drainage. (G) Complete embolisation was achieved via the posterior meningeal artery and posterior cerebral artery using Glubran, the middle meningeal artery using Onyx. (H) No recurrence was observed 3 months after complete embolisation. There has been a slight improvement in vision compared with before, and modified Rankin Scale score was 1.

DISCUSSION

Patient characteristics

In our study, most of the paediatric patients did not present with cardiorespiratory symptoms. However, asymptomatic cardiomegaly was found in 23.3% of the patients, with patients ≤1 year old being more likely to present (p=0.022).[4 19 20] Other symptoms, such as hydrocephalus and focal neurological deficits, were not observed to have significant age-related differences, unlike in other studies. Our results were similar to those in other major series with the most common DAVF location being the transverse/sigmoid sinus, torcular, superior sagittal sinus and cavernous sinus.[4 5 8] Other rare DAVF subtypes observed in our study included anterior cranial fossa DAVFs, isolated sinus DAVFs (figure 2) and intraorbital AVFs. In theory, DAVFs near the torcular may

have a worse prognosis due to involvement of the internal cerebral vein. In our study, 50% of torcular DAVFs had a poor prognosis, compared with 43% in Hett *et al*'s study.[4]

Treatment outcomes and prognosis

Intracranial DAVFs observed in children have distinct clinical presentations, angioarchitecture and clinical outcomes. The usual descriptions and adult classifications of DAVFs are not practically applicable.[1 2] According to a recent systematic review on paediatric DAVFs, 43.8% of the DAVFs were completely eliminated in 64 patients, with a complication rate of 29.7%.[3] Walcott *et al* reported 85.7% complete obliteration rate in seven patients, with good outcomes in all, but all of them were DSM and adult-type DAVFs.[10] Kincaid *et al* reported DAVF elimination in three/seven patients (43%), neurological deficit or developmental delay in three/seven patients (43%) and death in two/seven patients (29%).[5] Zaidi *et al* reported 11 paediatric DAVFs with 81.8% (9/11) complete obliteration rate and 18.2% (2/11) complication rate.[21] Hetts *et al* described 21 children treated with 38% complete obliteration rate, 70% good outcomes, 19% complications and 27% death rate.[4] Unfavourable evolution occurred in 50% of DSM and in 83.3% of infantile-type DAVFs. Except for one adult-type DAVFs who died from an unrelated leukaemia, all patients had favourable outcomes. Smajda *et al* reported 28 paediatric DAVFs in which endovascular treatment cured 68.2% of patients with DAVF with a clinical complication rate of 18.2% and a mortality rate of 13.6%.[7] Unfavourable clinical outcomes occurred in 66.7% of infantile-type DAVFs, 26.7% of DSM with DAVFs and 0% of adult-type DAVFs. The complete occlusion rate was 0%, 73.3% and 100% in infantile, DSM and adult-type DAVFs, respectively. In our study, 42 paediatric patients received treatment. Complete occlusion was achieved in 66.7% of the fistulas with 9.3% complication rate and 22.9% death rate. The complete occlusion rate was 35.3% (6/17), 75% (6/8), 88.9% (16/18) in infantile, DSM and adult-type DAVFs, respectively. Unfavourable clinical outcomes occurred in 57.1% (8/14) of infantile-type DAVFs, 16.7% (1/6) of DSM with DAVFs and 13.3% (2/15) of adult-type DAVFs. In comparison to DAVFs in adult,[22 23] paediatric DAVFs exhibited a significantly lower rate of complete occlusion, a higher rate of complications and a higher rate of mortality. Infantile DAVFs, in particular, had more treatment procedures, lower cure rates and a worse prognosis (table 3; figure 1). This was consistent with previous research findings.[4 7 8] In our cases, patients died more than 10 years after complete embolisation. As a result, even in patients who have had positive outcomes, long-term follow-up is required

Adult-type DAVFs

Adult-type DAVFs are found in all age groups, and almost all of these shunts are anatomically located at the cavernous venous plexus.[8] Isolated sinus DAVFs (primarily caused by thrombus) in the superior sagittal sinus and sigmoid/transverse sinus can also be classified as this type (figure 2).[4 18] Cure rate is high and postembolisation outcome is excellent.[4 7 8 10] Adult-type DAVFs are believed caused by trauma, thrombus and previous surgery, and so on.[8 11] In our study, 18 patients presented with adult-type DAVFs; 1 died as a result of an arterial infarction caused by embolic agent penetration across external–internal anastomoses into the cerebral parenchymal circulation, and 1 died for unknown reasons 16 years after complete embolisation.

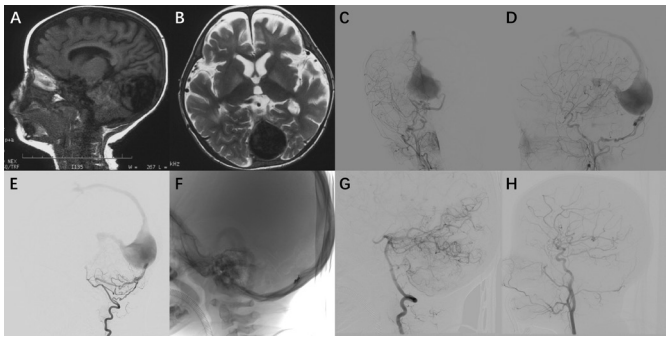

Figure 3 An infant presented with vomiting and seizure also diagnosed with congenital heart disease (ventricular septal defect). (A, B) Sagittal T1-weighted MRI (A) and axial T2-weighted MRI revealed a massive torcular herophili. (C–E) Common carotid artery and vertebral artery angiograms revealed a midline Cognard IIa+b/Borden II dural arteriovenous fistula with dural sinus malformation mainly supplied by bilateral occipital artery and dural branches of the left posterior inferior cerebellar artery. Note that both transverse sinuses have poor drainage, and the draining veins primarily involved the straight sinus-internal cerebral vein and the superior sagittal sinus-cortical veins. (F) Complete embolisation was achieved via the posterior inferior cerebellar artery using coils and Onyx. (G, H) There was no recurrence 21 months after complete embolization. He was asymptomatic, and no developmental delays had been found.

Dural sinus malformation

DSMs with DAVFs can be divided into two subtypes[8]: (1) DSM with giant dural lakes and slow flow mural arteriovenous shunting involves the posterior sinuses with or without torcular. Restricted outlets can often be seen due to thrombosis, occlusion or hypogenesis of the jugular bulb and sigmoid/transverse sinus (figure 3). (2) DSM involves the jugular bulb with otherwise normal sinuses but associated with a high-flow sigmoid sinus DAVF. The prognosis is also excellent with embolisation treatment.[4 7 8 10 24] DSMs are abnormalities resulting from disruptions in the embryological formation of the dural sinuses.[11] Yang *et al*[25 26] postulated that owing to the low level of flow, intrauterine spontaneous thrombosis of the shunts occurs with the persistence of a certain degree of ballooning of the sinus. The dural sinus thrombosis may play a role in DAVF formation.[27] In our study, eight patients presented with DSM. There were no deaths among the patients who were followed up on (six patients), but one patient was still

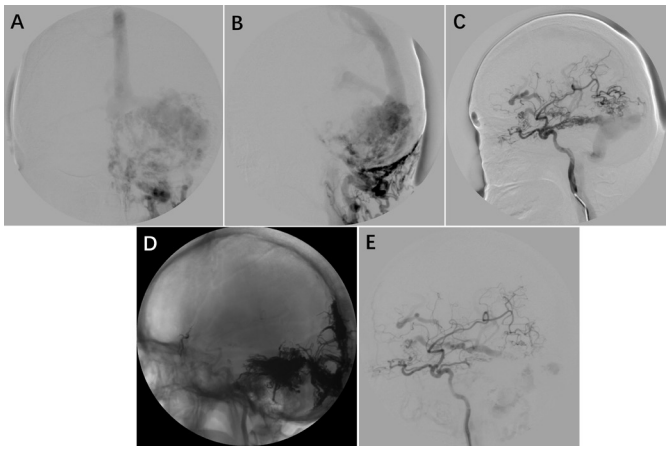

Figure 4 A paediatric patient in the age range of 10 presented with haemiplegia and seizure. (A–C) External and internal carotid artery angiograms revealed high-grade infantile type/multifocal (transverse sinus, anterior cranial fossa) dural arteriovenous fistulas mainly supplied by multiple dural and pial branches from the occipital artery, ophthalmic artery, meningohypophyseal trunk and middle cerebral artery, and so on. Due to bilateral transverse sinus obstruction, venous drainage primarily involved the straight sinus, the superior sagittal sinus and internal cerebral vein. (D, E) The blood flow in the arteriovenous fistula has significantly decreased after four treatment sessions, but new blood vessels continue to appear during each angiography examination and embolisation. The anatomic cure of the fistula was difficult to achieve, and the patient eventually decided to undergo regular follow-up. Unfortunately, the patient died 8 years after the last partial embolisation due to intracranial haemorrhage.

haemiplegic and mentally retarded 13 years after subtotal occlusion of the fistula due to hydrocephalus (mRS=3).

Infantile-type DAVFs

Infantile type/multifocal DAVFs are of high flow and low pressure type.[6 8 11] The sinuses are large, with no lakes and patent for a long time. They are thought to arise from widespread uncontrolled angiogenic processes, but the exact cause remains unknown.[28] The success rate of treatment and clinical outcomes are not satisfactory.[4 6–8 10] In some cases, it is difficult to achieve immediate cure of infantile-type DAVFs, and multifocality leads to recurrences with new dural or pial arteriovenous shunts opening near the previously embolised ones. The inability to achieve complete anatomical cure results in a high recurrence rate. Current embolisation techniques cannot treat complex multifocal dural shunts immediately because complete occlusion of all involved sinuses would result in severe venous outflow restriction.[5] We agree that partial embolisation to control symptoms and step-by-step treatment with preservation of normal venous drainage and reduction of cortical vein reflux are the best options (figure 4).[6 7] Grillner *et al* reported a case of multiple dural fistulas with large superior sagittal sinus.[29] Several cutaneous capillary malformations were observed on the child's extremities, and DNA analysis of the patient and father revealed a mutation in exon 21 of the RASA1 gene (c.2707C>T), which had previously been described as causing capillary malformation–arteriovenous malformation syndrome. Improved comprehension regarding their pathogenesis, combined with targeted antiangiogenic treatments, may have the potential to enhance the outcome of these patients. In our study, 17 patients presented with infantile-type DAVFs. Out of the 14 patients who were followed up, 6 patients died. They died at different time points after the last treatment: half a year, 1 year, 2 years, 2 years, 3 years and 8 years. The majority of them died from sudden intracranial haemorrhage with partial embolisation of the fistulas, while one patient died from multiple organ failure.

Similar to other large sample studies, paediatric intracranial DAVFs in our study had low rates of complete occlusion, poor outcomes and high rates of death, particularly in infantile-type DAVFs. DSM and adult-type DAVFs can now be easily cured by embolisation. However, treating infantile-type DAVFs is still difficult.

Study limitations

This study has some limitations because it is a single-centre, retrospective study. Self-report bias and the lack of core laboratory adjudication are two of these limitations. Compared with other studies, the incidence of hydrocephalus and macrocephaly in our study is much lower. We believe this is related to the generally older age of our patients.[3–5 7] Hydrocephalus and macrocephaly primarily occur in neonates and within the first year after birth.[4] The presentation of hydrocephalus differs significantly between those under 1 year old and those over 1 year old, with the above-mentioned symptoms being more prevalent in patients under 1 year old. In our study, only 10 patients presented relevant symptoms under 1 year of age, with 5 patients diagnosed with DAVF under 1 year old. Therefore, I believe this is the main reason why the proportion of hydrocephalus and macrocephaly was lower compared with other studies. The relatively small sample size of the paediatric DAVFs population in our series was also one of the main limitations due to the rarity, despite the fact that it is the largest single-centre study to our knowledge. Furthermore, due to the study's time span, not all patients were followed up on, which may have misjudged the final complete occlusion and survival rate. Finally, because our hospital is a tertiary referral medical centre for cerebral vascular disorders, the condition is more severe and complex due to referring bias.

CONCLUSION

Paediatric DAVFs are notable for their rarity, distinct clinical manifestations, low cure rate and poor prognosis. Most adult-type DAVFs and DSMs can now be effectively treated with embolisation, resulting in good outcomes and prognosis. However, there are still challenges in treating infantile-type DAVFs, and the prognosis is frequently poor. As a result, better understanding of their

pathogenesis, combined with targeted antiangiogenic treatments, has the potential to improve these patients' outcomes.

Contributors Conception and design: XS, YM, ZS, PZ. Acquisition of data: XS, ZS. Drafting the article: XS. Critically revising the article: YM, MY, PZ, HZ. All of the authors have read and approved the final manuscript. Gaurantor of overall content: MY, PZ, HZ.

Funding This study was funded by the National Natural Science Foundation of China (No.82101460).

Competing interests None declared.

Patient consent for publication Consent obtained from parent(s)/guardian(s).

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement Data are available upon reasonable request.

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