Dural Sinus Malformations (DSM) with Giant Lakes, in Neonates and Infants

Review of 30 Consecutive Cases

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Abstract

Background and Purpose

Dural Arteriovenous Shunt (DAVS) in children include Dural sinus malformation (DSM), infantile and adult types. They are rare and seldom reported. Our purpose was to highlight the angiographic features of the DSM sub group for prognosis of clinical evolution and outcome and to lay guidelines for management.

Methods: From a dedicated neurovascular data bank, there were 52 cases of arteriovenous dural shunts in children from 1985 to 2003. Of these, there were 30 patients with DSM, which we analysed the various angioarchitecture, presentation and neurological outcome. Children clinical status was evaluated and scored at admission and follow up.

Results

There was an overall male dominance of 2:1. Antenatal diagnosis was obtained in 8/30 (26.7%) cases. Mean age of diagnosis was 5 months. Mean age at first consultation was 8.7 months. No patient was diagnosed during childhood. The most common clinical presentations were macrocrania 76.7%, seizures 23.3% and mental retardation 23.3%. In 14/30 (35.7%) of the patients, the therapeutic decision was to manage conservatively; in 5/14 (30.7%) with predictable favourable evolution and in 9/14 (64.3%) with irreversible poor neurological out-

come. In the remaining 16/30 (53.3%) patients, endovascular treatment was performed. In 12/16 (75.0%) patients the neurological outcome was good, 3/16 (18.8%) patients had unfavourable evolution despite embolization. There was no morbidity mortality related to the procedures themselves. 1/16 (6.3%) patient was lost to follow-up. Overall 12/29 (45.8%) patients had an unfavourable neurological outcome with 11 patients dead and 1 with severe neurological deficit. In the surviving group of children, 17/18 (94.4%) have a good neurological outcome; in 10/18 (55.5%) the lesion is morphologically excluded.

Conclusion

DSM is rare disease with high mortality. They usually proceed to either total or partial spontaneous thrombosis before the age of 2 thus compromising normal cerebral venous drainage. DSM away from the torcular, good cavernous sinus, cavernous capture of sylvian veins, absence of pial veins, straight sinus or superior sagittal sinus (SSS) reflux and absence of jugular bulb dysmaturation represent factors of good prognosis. Such patients will highly benefit for endovascular treatment. In partial endovascular approach the aim being is to separate the brain drainage from DSM drainage. This will be achieved by the transarterial approach to the associated mural arterio-venous shunts (AVS) and by disconnecting the pial reflux by transvenous route.

Introduction

DAVS in the paediatric age group are rare, rarer than vein of Galen aneurysmal malformations¹, hence it is difficult to ascertain the incidence and prevalence. Morita et al (1995) reported a poor prognosis with overall mortality of 38%². There are few reported cases in the literature^{2,3,4,5,6}, most series published deal with adult group of patients. These papers did not exclude children, yet some are sometimes incorporated without specific attention.

DAVS in the pediatric population can be found as early as antenatally, in the first month of life or in the early infancy period, hence likely to correspond to a congenital malformation. They differ largely from adult ones that are likely to result from triggering causes rather than congenital ones¹.

Within paediatric DAVS three types can be individualised^{1,3}:

- Dural Sinus Malformations (DSM)
- Infantile type of DAVS
- Adult type of DAVS

DSMs include giant sinus lakes and single hole mastoid arterio-venous fistula (AVF) with jugu-

lar bulb diaphragm. From 52 consecutive cases of paediatric DAVS entered in our dedicated data bank the last 18 years we reviewed patients with DSM and giant lakes. Other forms of dural arteriovenous shunts (infantile and adult type of DAVS and DSM of the sigmoid sinus - jugular bulb diaphragm) were not included in this review. Despite the small size of this series, our purpose is to review the angioarchitectural features in an attempt to anticipate the clinical consequences of the various abnormalities seen and establish prognostic factors. Treatment goal and therapeutic timing were also reviewed.

Material and Methods

From 1985 to July 2003, we were referred 30 patients with DSM. The parameters prospectively tabulated included sex, date of birth, age at onset, age at first consultation, symptoms on admission, initial score, modality of treatment, number of sessions, materials used for embolization and follow-up outcome score (table 1). We use the 3 classic age groups to date the clinical onset: neonates (from birth to 30 days),

Table 1 **Dural sinus malformation with AV shunt (DSM)**

No	Sex	Age of Onset	Age of consult	Presenting features	Treatment	No of sessions	IS*	OS
1	M	birth	19d	Macrocrania, Cranial bruit	Heparin	0	-	0 Dead
2	F	23d antenatal	26d	Macrocrania, Cardiac failure, Hydrocephalus, ICH, Hypotonia	VPS 29d	A1-1m	-	4
3	F	birth antenatal	28d	Facial Haemangioma	0	A1- 1m	-	5 cure 28d
4	M	3d antenatal	28d	Ventriculomegaly, Seizures, ICH	0	0	-	0 Dead 1m
5	M	41d	43d	Generalised seizures, ICH, macrocrania, hypotonia	E1-arterial: 44d VPS: 49d E2-arterial: 4m E3-arterial: 8m	E1-1ped E2-3ped E3-3ped	2	3
6	M	4m	4m	Cranial bruit, ICH, Cerebral infarcts	0	0	1	0 Dead 5m
7	F	birth antenatal	3.5m	Cardiac failure, ICH, Hydrocephalus, Macrocrania	Heparin VPS: 9m	A1: 6m	1	0 Dead 10m
8	M	12d antenatal	16d	Macrocrania, Hydrocephalus	VPS: 13d	A1: 19d	-	1
9	M	6.5m	8 m	Psychomotor delay, Macrocrania, Raised ICT, IVH, Cardiac failure	0	A1: 9m	1	0 Dead 10m
10	M	1m	9m	Seizures, Raised ICT	0	A1: 3m	1	0 Dead 4m
11	M	7.3m	8.5m	Macrocrania, Mental retardation	0	A: 9m	1	0 Dead 10m
12	F	1m	5.3m	Facial Haemangioma	E1: 6m Heparin	E1:1ped A1: 1year A2: 2year A3: 4year	4	5 cure 4y
13	M	3m	3.8m	Generalised seizure, Bradycardia	Heparin	0	1	0 Dead 4m

No	Sex	Age of Onset	Age of consult	Presenting features	Treatment	No of sessions	IS*	OS
14	F	6m	1y 2m	Mental retardation, Macrocrania, Right orbital angioma	E1-arterial: 1y 5m E2-arterial: 1y 11m E3-arterial: 2y 4m E4-arterial: 2y 8m E5-venous: 3y 8m	E1: 3ped A1: 1y 8m E2: 3ped E3: 3ped E4: 3ped E5: sinus disconnection A2: 7y	3	4
15	M	4m	4.5m	Macrocrania, Dilatation of facial veins	E1-arterial: 6 m	E1: 1ped A: 1 y	2	5 cure 11m
16	M	6.5m	1y 7m	Macrocrania, Seizure, Mental retardation	E1-arterial: 1y 8m	E1: 2ped	1	No follow-up
17	M	1.5m	3.5m	Macrocrania, Seizure, Generalised hypotonia, IVH	Heparin VPS-3m 2d E1-arterial: 4.3m	E1: 2ped A1: 6m A2: 1y1m	2	5 cure 18m
18	F	1y 1m	1.5y	Cardiac failure, Mental retardation, Seizures, Macrocrania, Hydrocephalus	VPS-1y 3m E1-arterial: 1y 7m E2-venous: 1y 8m. E3-arterial and venous: 3y E4-arterial: 5y5m	E1: 3ped E2: sinus disconnection E3: 1ped sinus disconnection. E4: 2ped	2	4
19	F	28d	1.3m	Frontal cutaneous haemangioma, Macrocrania	Heparin E1-arterial: 3.3m E2-arterial: 7m	E1: 2ped E2: 2ped	4	0 Dead 9m
20	M	5m	9m	Macrocrania, Hydrocephalus, Raised ICT, Mental retardation	E1-arterial: 10m E2-arterial and venous: 13m VPS: 17m IVH E3-arterial and venous: 18m	E1: 3ped E2: 3ped Falcine sinus disconnection E3: 1ped SSS disconnection	2	0 Dead
21	M	5d	1y 5m	Orbital Haemangioma with anaemia, exophthalmus	E1-arterial: 1y 10m E2-arterial: 2y 2m	A1: 1y 9m E1: 3ped E2: 3ped	3	5 cure 3y
22	F	birth antenatal	4y 6m	Exophthalmus, Macrocrania	E1-arterial: 4y 8m	E1: 3ped	3	4
23	M	birth antenatal	4d	Macrocrania, CCF	0	A1: 3.8m	-	5 Cure 4m
24	M	6m	2y 11m	Macrocrania, Mental retardation Bruit, Exophthalmus	E1-arterial: 3y E2-arterial: 3y 3m	E1: 3ped E2: 3ped	3	4
25	M	birth antenatal	2.3m	Macrocrania, Hydrocephalus, ICH	0	A1: 3m	4	5 Cure 3m
26	F	4m	5m		E1-arterial: 5m	E1-1 ped	4	5 Cure
27	M	Birth	2y	Macrocrania, Bruit	E1-arterial: 3y	A1: 2y E1-2 ped A2: 4y	4	4 Cure
28	M	2y	4y	Macrocrania, Headache, Hydrocephalus, CCF, Bruit	E1-arterial: 5.1y E2-arterial: 5.2y E3-arterial: 5.4y E4-venous: 9.5y E5-venous: 9.6y E6-arterial & venous: 11.1y E7-arterial: 21y	E1-2 ped E2-3 ped E3-2 ped A1: 6y E4-sinus disconnection-coils Failed E6-2 ped -gel, particles E7-2 ped	4	4
29	F	Birth	2m	Macrocrania	Heparine E1-arterial: 1.4y	E1-2 ped	4	0 Dead
30	M	4m	8m	Macrocrania	0	A1: 1y 2m	4	4 Cure

*The IS score was not used in Neonates. IS: Initial score; A: Angiogram; OS: outcome score; E: Embolization; S: superior sagittal sinus; y: year; ICH: Intracerebral haemorrhage; m: months; ICT: Intracranial tension; d: days; VPS: Ventriculo-peritoneal shunt; ped: pedicle; IVH: Intraventricular haemorrhage; CCF: Congestive cardiac failure

Table 2 **Bicêtre Income and Outcome Score***

Score	Condition
5	Normal (N)
4	Minimal non-neurological symptoms (MS), not treated and/or asymptomatic enlargement of the cardiac silhouette
3	Transient neurological symptoms (TNS), non treated and/or asymptomatic Cardiac overload under treatment
2	Minor permanent neurological symptoms (MNS), mental retardation of up to 20%; Non permanent neurological symptoms under treatment; Normal School with Support and/or Cardiac failure stabilized with treatment
1	Severe neurological symptoms (SNS), mental retardation of more than 20% Specialized School and/or Cardiac failure unstable despite treatment
0	Death (D)

** does not apply to neonates*

infants (1 month to 24 months) children (2 to 15 years).

The clinical and neurological statuses of the patients were determined by paediatric neurologists which included the Brunet-Leizine and Denver neurocognitive tests. Infants were scored at admission and follow up with the Bicetre score¹ (table 2). Some of the patients had prior antenatal ultrasound (US) diagnosis, all patients were investigated by computed tomography (CT) and magnetic resonance imaging (MRI/MRA); before proceeding to angiography or embolization. All patients were evaluated neurological at 3, 6 and 12 months while undergoing treatment. Excluding the 1 patient lost to follow up, the post therapeutic follow-up period ranged from 3 to 84 months with a mean follow-up of 3.6 years. Angioarchitecture of 30 patients with DSM was analysed and correlated to the clinical scoring and evolution of the disease under treatment (table3).

The prominent angioarchitectural features selected were as follows:

1. Localisation of the DSM: DSM at the torcular or DSM away from the torcular.

Table 3 **Angioarchitecture of DSM with clinical score and clinical evolution**

No	AND	IS*	BD	CC	TI	Reflux	JBD	T	OS	Cause of death
1.	No	-	No	No	Yes	Yes	Yes	No	0	ICT
2.	Yes	-	No	No	Yes	No	Yes	Yes	4	
3.	Yes	-	No	Partial	No	No	No	Yes	5	
4.	Yes	-	Yes	No	Yes	Yes	Yes	Yes	0	ICH
5.	No	2	No	Yes	Yes	No	No	No	3	
6.	No	1	Yes	No	Yes	Yes	Yes	Yes	0	ICH
7.	Yes	1	No	Yes	Yes	No	No	No	0	ICH
8.	Yes	-	No	No	Yes	No	No	No	1	
9.	No	1	Yes	Partial	Yes	Yes	Yes	No	0	IVH
10.	No	1	Yes	No	Yes	Yes	Yes	No	0	ICT
11.	No	1	Yes	Yes	No	Yes	Yes	No	0	ICT
12.	No	4	No	Yes	No	No	No	Yes	5	
13.	No	1	No	No	Yes	Yes	Yes	Yes	0	ICT
14.	No	3	No	Yes	No	Yes	Yes	Yes	4	
15.	No	2	No	Yes	Yes	No	No	Yes	5	
16.	No	1	Yes	Yes	No	Yes	Yes	No	No f/u	
17.	No	2	No	Yes	No	Yes	Yes	Yes	5	
18.	No	2	No	Yes	Yes	Yes	Yes	Yes	4	
19.	No	4	No	Partial	No	No	No	No	0	ICH
20.	No	2	No	Yes	Yes	Yes	Yes	No	0	ICH
21.	No	3	No	No	No	No	No	Yes	5	
22.	Yes	3	No	Yes	No	Yes	No	No	4	
23.	Yes	-	No	Partial	No	No	No	Yes	5	
24.	No	3	No	Yes	No	Yes	Yes	No	4	
25.	Yes	4	No	Partial	No	No	No	Yes	5	
26.	No	4	No	Partial	No	No	Yes	No	5	
27.	No	4	No	Yes	No	Yes	No	Yes	4	
28.	No	4	No	Yes	No	No	No	No	4	
29.	No	4	No	Partial	Yes	No	No	No	0	ICH
30.	No	4	No	Partial	No	No	Yes	No	4	

*The IS score was not used in Neonates. AND-antenatal diagnosis; JBD-jugular bulb dysmaturation;
 IS-initial score at first consultation; OS-outcome score; TI-Torcular involvement; T-Thrombosis, ICH-intra cranial haemorrhage;
 BD-brain damage; ICT-Intracranial hypertension; CC-cavernous sinus capture

Table 4 Clinical manifestations*

Clinical features	%
Macrocrania	23/30 (76.7%)
Seizures	7/30 (23.3%)
Psychomotor delay	7/30 (23.3%)
Intracranial haemorrhage	8/30 (26.7%)
Brain damage	6/30 (20.0%)
Hydrocephalus	8/30 (26.7%)
Congestive cardiac failure	6/30 (20.0%)
Bruit cranial	5/30 (16.7%)
Facial veins dilatation	3/30 (10.0%)
Intracranial hypertension	3/30 (10.0%)
* Children may have more than one	

2. Complete, partial or absence of cavernous sinus capture (drainage of deep and superficial sylvian veins in the cavernous plexus).

3. Presence or absence of jugular bulb dysmaturation (post natal occlusion of the jugular bulb and retrograde thrombosis of the sigmoid sinus).

4. Persistence of a medial occipital sinus.

5. Presence or absence of pial veins, straight sinus or superior sagittal sinus (SSS) reflux (scheme 1).

The choice of treatment was either conservative with heparin or endovascular; be it transarterial or transvenous. The choice of embolization material was either glue or coil and in some cases both. Thirteen patients were not embolised due to either preexisting brain damage or severe initial score at the time of consultation.

Table 8 Analysis of cavernous capture

	Total cavernous capture	Partial carvenous capture	No carvenous capture
Favourable evolution	10 (71.4%)	5 (62.5%)	2 (25.0%)
Unfavourable evolution	3 (21.4%)	3 (37.5%)	6 (75.0%)
Lost to follow-up	1 (7.1%)	0	0
Total	14 (46.7%)	8 (26.7%)	8 (26.7%)

Table 5 Localisation of the Dural Sinus Malformation

	Torcular involvement	No torcular involvement
Favourable evolution	4 (28.6%)	13 (81.25%)
Unfavourable evolution	10 (71.4%)	2 (12.5%)
No follow-up	0	1 (6.3%)
Total no of patients	14 (46.6%)	16 (53.3%)

Table 6 Analysis of pial, straight sinus or SSS reflux

	Reflux	No reflux
Favourable evolution	6 (40.0%)	11 (73.3%)
Unfavourable evolution	8 (53.3%)	4 (26.7%)
No follow-up	1 (6.7%)	0
Total	15 (50.0%)	15 (50.0%)

Table 7 Analysis of dysmaturation of Jugular Bulb

	Dysmaturation jugular bulb	No dysmaturation jugular bulb
Favourable evolution	6 (37.5%)	11 (78.6%)
Unfavourable evolution	9 (56.3%)	3 (21.4%)
No follow-up	1 (6.25%)	0
Total no of patients	16 (53.3%)	14 (46.7%)

16 patients were treated endovascularly. The final clinical evolution was poor if the score was 0-2 and good if the score was 3-5 (table 3).

Results

Demography

A male dominance was noted (1.9: 1). the oldest patient at diagnosis was 2 years, mean age of 5 months in the series. Mean age at first consult in our centre was 7 months for a maximum at 4 years.

Antenatal Diagnosis (figure 1)

8 (26.7%) patients were diagnosed antenatally during routine US; half of which were of torcular type of DSM, for these the M:F ratio was 1:1. Six patients (75.0%) had a favourable outcome and 2(25.0%) with unfavourable outcome one of which presented with brain damage.

Clinical Presentation

The most frequent clinical presentation was macrocrania 76.7.0%. Seizures psychomotor delay and Intracranial haemorrhage (ICH) each were present in 23.3%. Brain damage was noted in 20%; while hydrocephalus in 24.0% (table 4). The other clinical presentations are congestive cardiac failure (CCF), cranial bruit, facial vein dilatation and intracranial hypertension (ICT) the latter being associated to macrocrania.

Angiographic Findings

DSM involving torcular was noted in 14 (46.6%) patients (table 5). Pial, straight and superior sagittal sinus reflux was present in 15 (50.0%) (table 6). Dymaturation jugular bulb was demonstrated in 16 (56.0%) patients (table 7).

In our study, total bilateral cavernous sinus capture was seen in 14 (46.7%), partial cavernous sinus capture was present in 8 (26.7%). In 8 (26.7%) cases, no cavernous sinus capture was noted (table 8).

Table 9 Clinical evolution patients

	Embolized	Unembolized
Favourable evolution	12 (75.0%)	5 (35.7%)
Unfavourable evolution	3 (18.8%)	9 (64.3%)
Lost to follow-up	1 (6.3%)	0
Total	16 (53.3%)	14 (46.7%)

Treatment Option and Evolution

16 (53.3%) patients were embolised, 12 (75%) with glue via transarterial approach, 4 (25%) were treated with glue and coils via transarterial and transvenous approaches. There was no morbidity mortality related to the procedures themselves. The clinical evolution of 16 embolised cases was good in 12/16 (75.0%) (of which 8/12 (66.7%) cured) and poor in 3/16 (18.8%) where all died despite embolization. There was 1/16 (6.3%) patient lost for follow-up. The clinical evolution of 14 (52%) conservatively treated patients was good in 5 (35.7%) where the favourable out-

Table 11 Overall results

Clinical situation	No of patients
Favourable clinical evolution	17/29(58.6%)
Favourable clinical evolution with morphological exclusion	10/17(58.8%)
Unfavourable clinical evolution	12/29(41.4%)
Unfavourable clinical evolution with death	11/12(90.9%)
No follow-up	1/30(3.3%)

Table 10 Final clinical score

	Score 0	Score 1	Score 2	Score 3	Score 4	Score 5
Initial *	0	7/24 (29.2%)	5/24 (20.8%)	4/24 (16.7%)	8/24 (33.3%)	0
Outcome**	11/29 (37.9%)	1/29 (3.4%)	0	1/29 (3.4%)	9/29 (31.0%)	7/29 (24.1%)
*The IS score was not used in Neonates.; **one patient was lost to follow-up.

come was expected without treatment, (4 patient with spontaneous thrombosis) (figure 2-3). The evolution was poor in 9 (64.3%) where no acceptable therapeutic goal could be set, all died (table 9) (figure 4).

The analyses of the final clinical results are as in (table 10). Initial score was good (score 3-5) in only 12 (50.0%). Good outcome score was noted in 17/19 (89.5%) surviving children. The overall outcome are as in table 11. Favourable clinical evolution with morphologic exclusion was noted in 10/17 (58.8%). Conversely 11/12 (90.0%) with unfavourable evolution died.

Discussion

Epidemiology

During the same period from 1985-2003, there were a total of 1566 patients with intracranial AVMs entered in the same data bank: 946 adults and 620 children. In the paediatric age group there were 317 Vein of Galen malformations (VGAM) and 303 non-galenic malformations. There were 52 DAVS of which there were 30 DSM with giant lakes involving the torcular and/or adjacent sinuses. Therefore our study group represents 1.9% of the total

Figure 1
Antenatal MRI
diagnosis of DSMs
with (A, B) and
without (C, D)
evidence of intra
luminal thrombosis.

AVM (for a total of 1618 if we include DSM), 4.8% of the paediatric population, and 57.7% of the DAVS in paediatric group seen during the same period of time.

There have been few reports to date that addressed the entity of dural arteriovenous shunts (DAVS) in the paediatric age-group^{2,3,4,5}. In this group, neonates and infants constitute a special subgroup¹. Nosologic and clinical differences between neonates, children and adults are found in the various types of DAVS, as each population has its own compliance and vulnerability. DAVS has been reported to have

a mortality rate of 38.0% taking all ages together, 67% in the neonatal subgroup, in Morita series³ and 37.9% in our followed up patients.

DSMs generally reveal in neonatal and infancy periods although symptomatic on the average at 5 month of age (scheme 2).

26.7% of our DSM case were diagnosed by antenatal US, hence higher the chance of early diagnosis in DSM, provided the Ultrasonographer is aware of this rare disease. In our series of VGAM 28.0% were diagnosed antenatally.

Figure 2 (Case 23) A-C) Antenatal MRI diagnosis of DSM. D-F) post natal evidence of intra luminal thrombosis. G-I) complete remodelling.

Clinical Evolution

Early symptoms can be cardiac failure (usually mild and unfrequent) in neonates, coagulation disorders (consumptions syndromes), moderately increased intracranial pressure (with irritability, macrocrania, neurocognitive delay and seizures) in infants^{1,3,8}. Hence, DSMs with their mural AVS are different from neona-

tal and infant AVS as the shunts in DSMs seem a secondary phenomena at the level of the sinus, usually with low flow characteristics. Among the six patients with brain damage, five died and 1 was lost to follow up. The cause of death was ICH in 2, IVH in one and uncontrollable raised ICT with tonsillar prolapse in the remaining two.

Figure 3 A-B) Neonatal MRI diagnosis of DSM away from the torcular. C-D) post natal spontaneous thrombosis.

In four patients who presented with no brain damage, diagnostic angiography showed no cavernous capture with involvement of torcular, the evolution was bad in three (two dead) and good in one. The patient with good evolution did not have a good delayed venous phase images to evaluate the cavernous capture properly (figure 5), however alternate pathways were likely to be sufficient (patient no 2). In this group of patients, the absence of cavernous capture, and the involvement of torcular gave rise to a unfavourable neurological evolution

related to spontaneous thrombosis of the torcular. Here the timing of diagnosis and treatment is crucial, to avoid early torcular thrombosis before the cavernous sinus capture occurs.

Seven patients presented without brain damage, with partial or no cavernous capture, and the DSM seated away from the torcular. The follow-up showed six patients with good evolution (six cured) and one with poor evolution. This patient with poor evolution had progressive increase in size of her malformation for nine months and later expired due to intracra-

Figure 4 (Case 19) A-C) neonatal MRI showing torcular DSM. D-F) 7 months FU MRI shows multiple cavernomas with haemorrhagic changes in the right periventricular region and enlargement of the DSM to the Right transverse sinus.⁸

nial haemorrhage due to associated cavernomas⁸ (figure 4).

Other patients had better evolution. Lateralisation of the DSM, or a location away from the torcular on the SSS allowed the brain to drain through the contralateral sinus or through Labbe's veins downstream the DSM. In three patients of this group, spontaneous cure with remodelling of the sinuses was observed (figure 6).

Embryology and Pathophysiology

Anatomically two types can be seen (only the first one was studied here):

- DSM involving the adjacent posterior sinuses, with giant pouches and slow flow mural AV shunting. Partial thrombosis of the sinus may occur and can be also be observed in utero. The dural sinus pouch at birth communi-

cates with the other sinuses and drains normal cerebral veins.

- DSM of the jugular bulb malformation with otherwise normal sinuses appears as a sigmoid sinus-jugular bulb "diaphragm" and is associated with a petromastoid-sigmoid sinus high flow AVF which is usually a single hole type¹. These ones have a usually benign course, remaining asymptomatic for a long time and being incidentally discovered. Their treatment by embolization is technically easy and end up with complete exclusion of the shunt and favourable neurological outcome.

According to Okudera⁷ the fetal changes of the sinuses, goes through a apparent relative ballooning of the transverse sinus, which occur at 4-7 months intrauterine which then progresses gradual remodelling to post natal stage after 1 year of age. Thus the posterior sinus DSMs

Figure 5 (Case 14) A-D) Mural AVS from the R. intra cavernous ICA. No deep vein seen, bilateral cavernous sinus capture, posterior fossa congestion. Jugular veins patent bilaterally downstream jugular foramina.

have been thought to correspond to an abnormal perinatal persistence of the sinus ballooning. However, this does not explain why the so called normal ballooning is not seen in the antenatal period in normal cases. Cases of DSM are by definition, associated with the uncontrolled development of posterior sinuses including transverse, sigmoid sinus and/or confluence of sinuses. Hence, DSM is a disease of the sinuses development instead of the embryolog-

ical non development. This accounts for the progression of the disease with sinus wall overgrowth, abnormal development of epidural confluent of venous spaces leading to segmental giant lakes followed by secondary thrombosis of the spaces and subsequent remodelling if the venous drainage of the brain can be rerouted. In one case DSM ongoing increase in size was associated with the appearance of haemorrhagic cavernomas⁸ (figure 4) No hereditary

Figure 5 E-G) Mural AVS from the ascending pharyngeal and middle meningeal arteries; - Torcular involvement - Straight sinus and SSS reflux - Left Jugular bulb dysmaturation. H) retrograde injection of the straight sinus prior to its trans venous disconnection. Note the opacification of the cavernous drainage of the deep system.

vascular disease is associated with DSM, in particular HHT. There is no familial history of DSM in our group of patients, indicating the absence of germinal implication in DSM.

Associated slow flow multiple AV shunts are consistently noted within the wall of the dural wall of the DSM; they add to the hydro-venous restriction by venous congestion of the brain, as the brain at birth has to drain through the diseased sinus. This added constraints to the normal brain venous drainage will persist until cavernous capture of the sylvian veins provides

an alternate outlet towards the ophthalmic veins or pterygoid venous plexuses^{1,3}. Early and rapid spontaneous thrombosis of the DSM lake and secondary dysmaturation of the jugular outlets further compromise cerebral venous drainage and subsequently lead to venous infarction and lethal intraparenchymal haemorrhage. As long as the venous outlets are patent, the clinical manifestations remain contained.

The lateralised DSMs away from the torcular have better chances of favourable outcome as there is one normal sinus to allow the brain to

Figure 5 I-M) Following trans venous occlusion of the straight sinus, Left transverse and SSS with coils (I), few AVS were embolised leading to substantial reduction in flow into the malformed sinus (J-M)

drain. However it is important to have the ipsilateral cerebral hemisphere drainage in an alternate pathway either by cavernous capture, or by a persistent medial occipital bypassing the thrombosed sinus into the ipsilateral jugular vein or into the contralateral sinus via the SSS.

Treatment strategy

The therapeutic options thus depend on the individual cases angioarchitecture and state in the maturation or dysmaturation process.

Figure 5 N-Q) 4 years FU angiogram shows remodeling of the cerebral venous drainage. Note the bilateral cerebellar DVA draining into the Left petrous vein and subsequently into the cavernous sinus and downstream the bulb occlusion into the Left jugular vein.

When there is partial or no cavernous capture and no pial reflux, there is an option of treating with heparin and embolising the shunts with glue expecting cavernous capture to take place with minimal or no consequences for the hydro venous equilibrium of the maturing brain and granulations. If there is significant shunt causing pial reflux embolization is necessary to prevent venous hypertension and cerebral ischemic damage.

The goal is to reduce to pial reflux although the shunts may preserve the patency of the sinuses. If the mural AVS are completely occlud-

ed, the sinuses are likely to thrombose, resulting in absence of outlet for the brain leading to poor evolution.

In 7 patients without brain damage, bilateral cavernous capture and no jugular bulb dysmaturation, there was good evolution in six (three cured). One patient died from haemorrhagic complication following ventriculoperitoneal shunt. A patient presented without brain damage, bilateral cavernous capture, jugular bulb dysmaturation, but no pial, straight sinus or SSS reflux. This patient had his mural AVS embolised with later good evolution. Five patients

Figure 6 (Case 15) Remodelling following trans arterial embolization of mural AVS.

presented without brain damage, bilateral cavernous capture, jugular bulb dysmaturation and reflux into pial veins, straight sinus or SSS. These patients were embolized transarterially for their AVS and transvenously by coils) to disconnect the pial vein openings (figure 5). Four patients had good subsequent evolution. One of them had stable evolution for 18 months and later had intraventricular haemorrhage due to persisting pial reflux.

There has been no attempt to surgical sinus bypass or stenting of the jugular bulb, both being known (although unpublished after initial morphological good results) to rapidly thrombose.

Conclusions

DSM is a rare disease of the posterior sinuses which may start antenatally. They correspond to a distinct entity within the DAVS and even in the pediatric subgroup of DAVS. DSM is not a hereditary disease.

Following antenatal diagnosis, we suggest MRI of brain to see the location of the DSM and disclose already existing brain damage. If the DSM is located on the midline and large early angiogram is recommended to assess the anatomy of cerebral veins. If the DSM is moderate in size, follow up MRI at two months interval should verify its evolution and that of the maturing brain.

Angiogram is to be done at 4-5 months in all

the DSM patients to have a prognostic evaluation at a time when cavernous sinus may have taken place. Further management of conservative, heparinisation (low molecular) or endovascular treatment is decided and performed at the same time. Brain damage, DSM involving torcular and absence of cavernous capture, are pejorative features.

DSM away from the torcular, presence of bilateral capture cavernous and absence of jugular bulb dysmaturation are favourable findings. The treatment options must be adjusted to each situation. Partial embolization of the AVshunts and/or disconnection of venous drainage to the brain will protect the brain and allow later for the exclusion of the lake if its thrombosis has not occurred spontaneously.

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