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Congestive cardiac manifestations from cerebrocranial arteriovenous shunts*

Endovascular management in 30 children

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Abstract. Since 1984 we have been involved in the management of 30 children who had cardiac manifestations secondary to cerebrocranial arteriovenous shunts. Aneurysm malformation of the vein of Galen was the most common vascular lesion observed (73% of cases). In 77% of the patients the cardiac symptoms were the main presenting complaint. Medical treatment and/or endovascular therapy were indicated, depending on the age of the patients and the severity of the cardiac manifestations. Following embolization, the cardiac symptoms resolved (73%) or improved (18%) in 1 or 2 sessions. Mortality in the embolized group was 9%, and transient nonneurologic morbidity occurred in one case. Overall mortality, including four patients rejected for embolization, was 20%. These results compare favorably with medical and/or surgical management, alone or combined. The technique, challenges, indications and contraindications of endovascular therapy are discussed. Embolization represents an effective adjunct treatment to control, improve or cure the congestive cardiac manifestations caused by cerebrocranial arteriovenous shunts.

Key words: Cardiac failure – Cerebral arteriovenous malformation – Embolization – Neonate – Pediatric angiography – Vein of Galen malformation

Cerebrocranial arteriovenous shunts (AVSs) are infrequent causes of congestive cardiac failure (CCF). When CCF is suspected following clinical examination including cranial auscultation, it can easily be confirmed by transfontanel ultrasound [13]. However, most of these infants are first considered to have congenital heart disease [5, 10, 12, 13] and are sometimes subjected to cardiac angiography [10, 12, 13].

Cardiac manifestations secondary to cerebrocranial AVS are extremely variable in intensity: from severe heart failure resistant to medical treatment to well-tolerated, mild cardiac overload. In the latter, the only sign is an asymptomatic cardiomegaly demonstrated on routine chest radiograms in children with neurological symptoms.

When presenting in a newborn, the prognosis of severe heart failure from a AVS of cranial origin is very poor with 100% mortality [6, 7]. However, in recent years, the use of endovascular therapy in newborns and infants has significantly changed the classically poor outcome of these patients [9]. Arterial embolization, although technically challenging in babies weighing only a few kilograms, allows dramatic improvement of cardiac function.

The purpose of this paper is to report our experience with embolization in CCF of cerebrocranial origin and to discuss the best timing for the intervention as indicated by the patient's symptoms.

Patients and methods

Since 1984, one of us (P.L.) has been involved in the consecutive mangement of 30 children with cardiac symptoms secondary to cranial AVS. No referred case has been excluded since that time. The age at the time of diagnosis was: 0–1 month in 19 cases, 1–12 months in 7 and 1–15 years in 4. There were 18 male, and 12 female patients. In all of them, by carefull cardiac examination (including ECG and ultrasound), we sought a cardiac anomaly responsible for the symptoms. In 23 (77%) patients, clinical CCF (with tachycardia, polypnea, hepatomegaly) was the main complaint. In the remaining 7 (23%), macrocephaly or hydrocephalus was the prominent manifestation, mild heart failure (stable with medical treatment) or an incidental cardiomegaly being discovered by complementary studies.

The type of vascular lesions responsible for the cardiac symptoms were: vein of Galen aneurysm malformation (VGAM) (22 patients), cerebral arteriovenous malformations (2 patients), dural arteriovenous fistulae (2 patients), facial capillary hemangioma (2 patients), cerebral arteriovenous fistula (CAVF) (1 patient) and facial arteriovenous fistula (1 patient).

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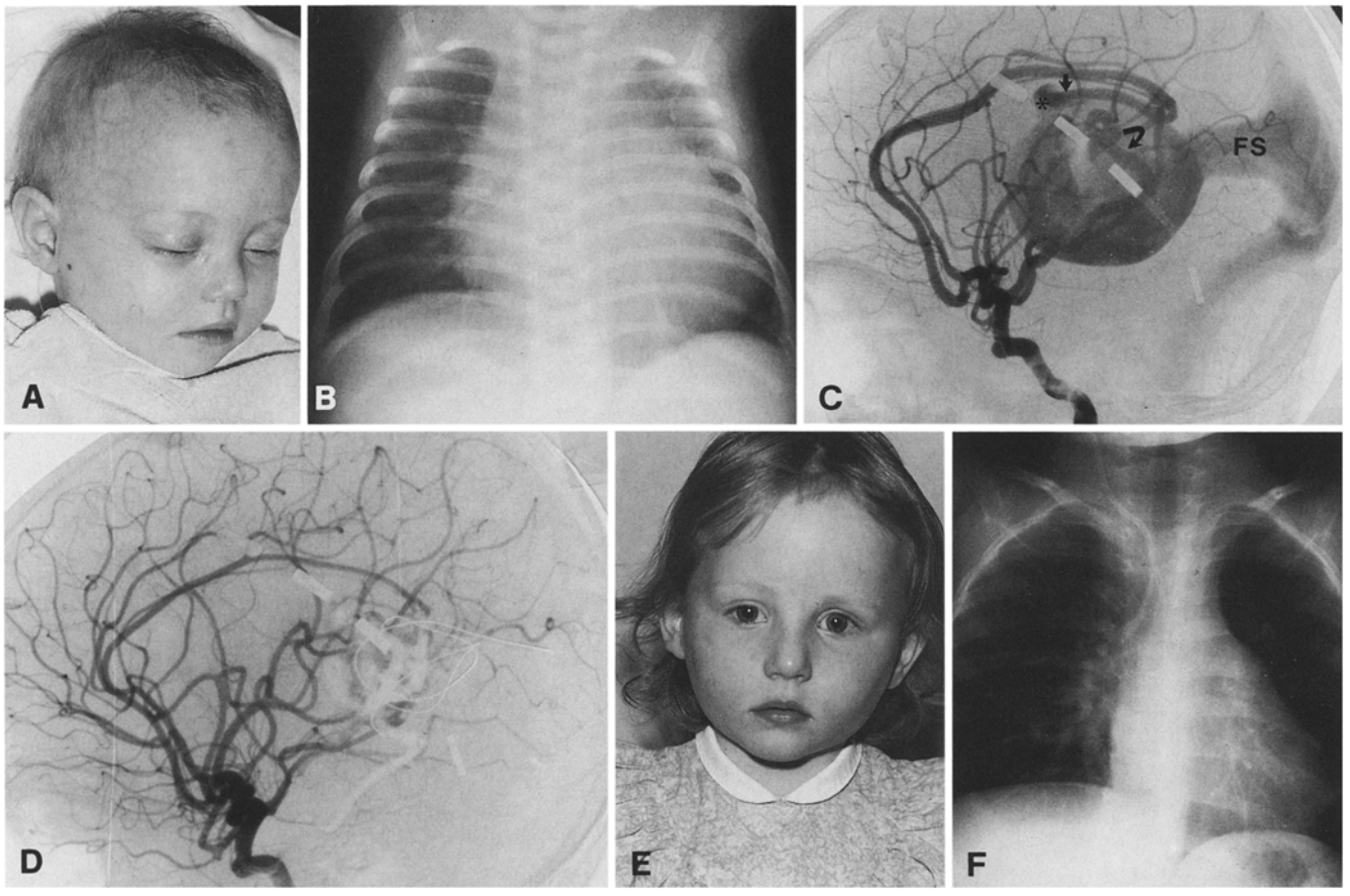

Fig. 1. **A** Newborn who presented with severe heart failure and secondarily collateral venous circulation of the face and scalp. **B** Chest film shows cardiomegaly and increased pulmonary circulation. **C** Carotid arteriography (lateral view) shows a choroidal vein of Galen aneurysmal malformation with blood supply from the subfornical branch of the pericallosal artery (*arrow*) and posterolateral choroidal artery (*curved arrow*). An arteriovenous shunt (AVS) (*asterisk*) in the wall of the ectatic vein drains towards the torcula through the falcine sinus. **D** Carotid arteriography after combined arterial and venous embolization. No remaining AVS is seen. Note the reduction in caliber of the pericallosal and subfornical arteries in comparison to **C**. **E** The child at present is asymptomatic without medication (4 years after **A**). **F** Present chest film. Normal cardiothoracic index and pulmonary vessels

All the patients with symptomatic heart failure were treated with duretics and digitalics at the time of referral. Arterial embolization of the vascular lesion was indicated in 24 patients and actually performed in 22 (57 arteries in 42 sessions). At the time of first embolization, this population consisted of 5 newborns, 11 infants and 6 children. Among them, 9 weighed less than 5 kg, 9 between 5 and 10 kg, and 4 weighed more that 10 kg. In 10 patients embolization was done on an emergency basis.

Endovascular therapy was performed in all cases by a femoral approach with the Seldinger technique through a 4 F. sheath. At the time of referral, two newborns had one patent femoral artery. In both cases the right femoral artery was impaired following diagnostic angiograms performed in another institution (by the percutaneous, and cut-down approach, respectively). Low-osmolar contrast material was employed in all cases. Isobutyl cyanoacrylate or *N*-butyl cyanoacrylate was used to embolize all vascular malformations or CAVFs, whereas capillary hemangiomas of the face were embolized with micro particles of polyvinyl alcohol foam.

Results

Embolization was performed in 22 patients. CCF completely resolved in 16 (73%), cardiotonics and diuretics being discontinued within a few days following embolization. In four patients, (18%) endovascular therapy resulted in significant clinical improvement. In these patients medical treatment could be tapered down but not discontinued. A 9% (2 patients) mortality occurred in the embolized group: a 3-kg baby died of massive gastrointestinal and cerebral hemorrhages 1 week after partial embolization of a VGAM. The other death in our series occurred in a 3-week-old newborn with a brain stem and a cerebellar arteriovenous malformation. This infant had major hemodynamic disorders throughout the intervention; postoperative ultrasound showed a cerebral hemorrhage. The overall mortality in our series of 30 patients, including both the embolized and the non-embolized, was 20%.

A 4.5% (1 patient) morbidity was noted in a 6-month-old male infant with severe heart failure from a giant facial capillary hemangioma. It was resistant to steroid therapy, thus treated by emergency bilateral maxillary and facial embolization. Improvement in cardiac function was rapidly obtained but fever without infection and pain after the procedure lasted a few weeks. Skin

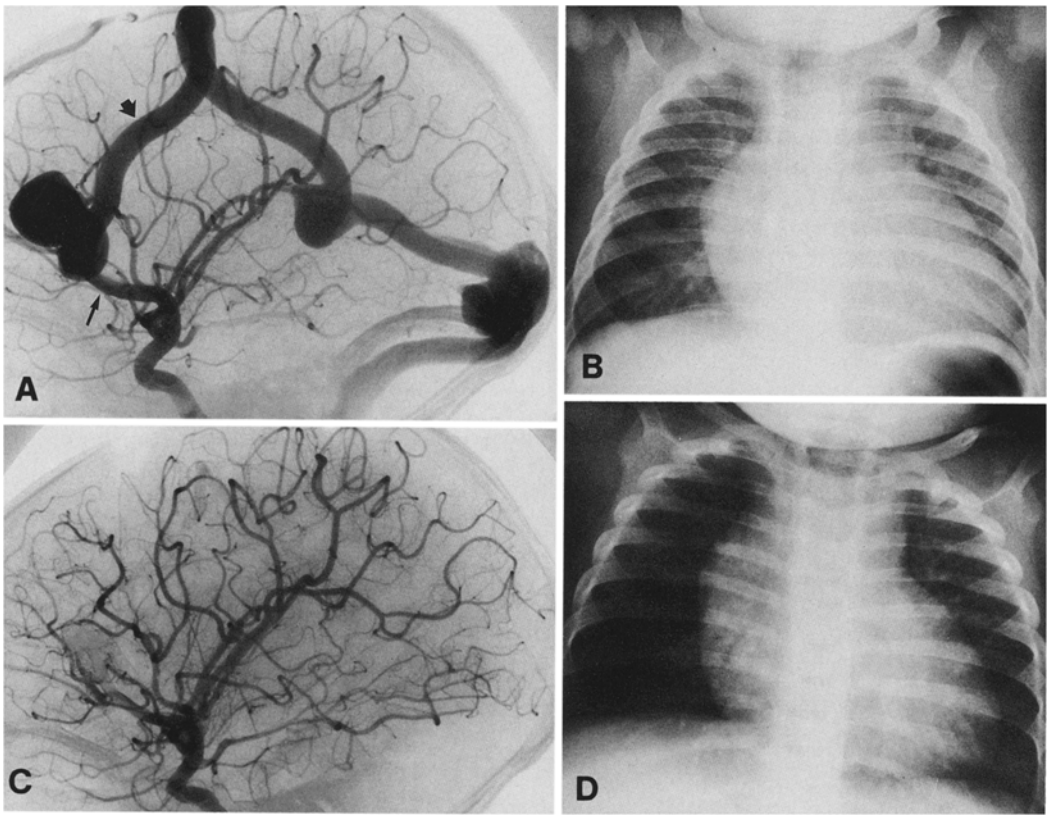

Fig. 2. **A** Carotid angiography in a 8-month-old girl with mild heart failure shows a cortical AVS localized in the anterior cingulate gyrus. Arterial supply from the anterior cerebral artery (*thin arrow*) with venous drainage through an internal frontal vein (*solid arrow*) towards the vein of Galen and straight sinus. **B** Chest film shows cardiomegaly with increased pulmonary circulation. **C** Carotid angiography after arterial embolization reveals occlusion of the AVS. **D** Control chest film 1 month after embolization reveals reduction of the cardiomegaly

necrosis was noted secondarily (2 × 2 cm) on both cheeks. Healing occurred spontaneously in a few months without skin grafting.

There was no neurological morbidity related to embolization in any of the other cases. No femoral complication occurred during or after the procedures. In all cases the femoral approach was possible by percutaneous puncture. In one case, the puncture was done under doppler guidance due to an "unpalpable" femoral artery. We had no permanent technical failures, although in 4 cases endovascular therapy could not be achieved on the first attempt. These patients were successfully embolized some days later.

The results of our management are shown in relation to three different practical choices made following the answering of two major questions: Should we treat? Can we wait? In group 1 (therapeutic abstention), endovascular therapy was rejected in 2 patients because of severe brain damage demonstrated by ultrasound and computed tomography (CT). The remaining 2 cases were rejected because of technical limitations: the arterial feeders of the lesion were felt to be inaccessible for significant and safe embolization. All the patients in this group died.

Group 2 consisted of 7 neonates, whose cardiac failure was responsive to medical treatment with diuretics and/or digoxin. In these cases, embolization was postponed until the infants were 6–8 months of age. The rationale was that if there was no urgent need to control the shunt, intravascular navigation and embolization would be easier in a 5-, to 6-kg baby than in a 2.5, to 3.5-kg baby. This delay is acceptable if one assumes that the CCF remains responsive to medical treatment and that the neurological examination and thriving are normal. In the meantime, these patients are periodically examined clinically and monitored by CT studies to disclose any sub-clinical abnormality. As the result of this careful follow-up, in three cases the embolization was anticipated and therefore performed before the elected time; in two cases the CCF became resistant to medical treatment; in the remaining, a mild cerebral atrophy was demonstrated on follow-up CT. These 3 patients are presently free of cardiac symptoms and one neurologically normal. The other 4 patients in this group grew up with stable cardiac function under medical treatment. At the elected time, 2 of them were embolized and their lesions anatomically excluded. The remaining 2 are still waiting for elective embolization.

Group 3 comprises 19 patients in whom immediate embolization was chosen as an adjunct to medical treatment following careful analysis of the cerebral tissue status [by ultrasound, CT, or magnetic resonance (MRI)]. In 10 of them, severe heart failure resistant to medical treatment required urgent endovascular therapy (Fig. 1). In the other 9 patients, all older than 6 months, the cardiac status did no represent the primary cause for immediate embolization (Fig. 2). In 2 cases, the parents refused endovascular treatment; both are lost to follow-up.

Discussion

CCF secondary to AVS has been well documented in numerous reports [1, 7, 9, 11, 14]. It represents an infrequent clinical finding in vascular lesions of the head.

From 600 referred cases of cerebrocranial vascular lesions, only 30 (5%) presented with cardiac manifestations. However, in our pediatric population this percentage rose to 19%. Some types of isolated high-flow fistulae in children amazingly are seldom associated with cardiac manifestations: vertebrovertebral and external carotid fistulae. VGAMs, on the contrary, are frequently associated with cardiac manifestations. They represent 73% of our population with CCF or cardiomegaly of cranial origin. Unfortunately, this association is frequently not suspected. Cardiac angiography is often performed to find a congenital heart disease [10, 12, 13]. Angiocardiography is unnecessary, and may transiently or permanently impair the femoral vessels. In fact, the cardiac manifestations are not specific to their cranial origin, but are common to CCF of other causes. Cyanosis may exist in the presence of right to left AVSs, as atrial communication or ductus arteriosus [5, 11, 13]. This persistance of a fetal type of circulation should not be regarded as a true cardiac anomaly since it reflects right atrium volume and pressure overload.

In our series, severe heart failure in a newborn was always secondary to an intracranial vascular lesion; galenic, cerebral or dural AVSs can equally give rise to severe CCF [4, 8]. In addition, an intracranial AVS will produce cardiac failure only at a very young age (1–19 days in our series). The older the child, the lower the changes are of cardiac manifestations and the milder they will be. Mild heart failure or simple cardiomegaly are observed as additional symptoms in infants whose chief complaints are macrocephaly or other neurological manifestations [11]. In these cases, the etiologic diagnosis occurs later, usually after 6 months of age.

The prognosis of severe CCF of cranial origin in newborns or infants has classically been considered very poor [13]. Previous authors report a mortality between 63% and 90% under medical or surgical treatment [7]. In the pediatric population, this poor outcome has improved significantly with modern endovascular techniques, with an overall mortality of 20% in our series. Similar results were reported by Berenstein and Epstein on a population with VGAMs [3].

Treatment of CCF with giant capillary hemangiomas of the face is different. Symptoms start with the proliferation phase of the lesion at 4 to 12 months of age. The objective here is to exclude the lesion from the general circulation and to gain time to allow spontaneous regression to occur [8].

Arterial embolization presents some distinctive features when performed in newborn and infants. The small size of femoral arteries requires the use of small catheters to perform angiographic examinations and embolizations. Following a rigorous protocol and decision process, the procedure should be performed as soon as possible to reduce the catheterization time. Post-intervention, femoral patency is essential to allow staged embolization if needed, and future normal growth of the lower limbs. Contrast material is a limiting factor in these babies weighing only a few kilograms because of the danger of volume overload. In our series, only small amounts (4–6 ml/kg body weight) of low osmotic contrast material were used with perfect tolerance and no worsening of the cardiac status.

In all cases, the general anesthesia necessary to perform the procedure constitutes another challenge. Tracheal intubation, assisted ventilation, pulse oxymeter, permanent ECG and noninvasive, automatic pressure monitoring are mandatory. The anesthesic medications (barbiturates, curare, isoflurane, fentanyl) are potentially dangerous in patients being treated with digitalics[1] and diuretics.

Accurate hemodynamic balance should be maintained during the procedure. Coaxial system perfusion and contrast administration may worsen the cardiac status. Arterial hypotension should be avoided during anesthesia because of its deleterious effects on brain and myocardial perfusion [6]. In spite of these limiting factors, endovascular therapy, if performed by a well-trained, multidisciplinary team, offers a very low morbidity [9], few technical failures and dramatic efficiency.

The goal of arterial embolization in newborns with severe heart failure is to decrease the flow of the cranial AVS. Eventually, the volume and pressure overload to the right atrium will be reduced, allowing stabilization of cardiac function. Complete anatomical cure of the cerebral lesion is not mandatory at this stage. Beneficial changes in cardiac hemodynamics and symptomatic relief rapidly follow partial embolization. However, we do not know what percentage of the shunt should be occluded to obtain a significant clinical result. Empirically, at least 30% of the lesion must be controlled in one procedure. Subsequent oral alimentation would allow normal thriving and weight gain until the child is older. Subsequent embolization(s) to achieve complete anatomical exclusion can then be performed some months later when the patient's general condition has improved. However, in some cases complete occlusion of the shunt is achieved in one session. In such a case, hospitalization in an intensive care unit following embolization is advisable. Positive pressure ventilation, diuretics, and sedation during a 36-h period are recommended to protect the heart from the rapid increase in overload following an abrupt occlusion of the shunt. In the presence of a fall in arterial blood pressure, an infusion of dobutamine (7.5 μg/kg body weight per min) is used.

In view of the technical ease of endovascular procedures in babies weighing 5 or more kg, we think that embolization does not need to be done in a newborn whose heart failure responds to medical treatment. In such cases, we suggest periodic clinical follow-up and MRI examinations to survey neurologic development. Occurrence of a warning sign or symptom should prompt embolization. If not, pre-therapeutic angiography is proposed at 6–8 months of age to determine the flexibility of an endovascular procedure. In these cases, we think there

[1] Although widely used, we believe that digoxin is not indicated because of its negative chronotropic effect, since tachycardia is a normal adaptative response of the heart in cases of congestive failure without impairment of systolic function. This problem will be addressed in a future report

is no indication for a "diagnostic" angiogram in early infancy since CT and MRI provide all the necessary diagnostic information.

Although the purpose of the present paper is to report the results we achieved in the management of the cardiac manifestations, we remain concerned about the neurological future of these infants.

Conclusion

The traditional poor prognosis of congestive heart failure from cranial origin in early childhood must be re-evaluated. Management of these patients by the endovascular team will help to select the most appropriate treatment. Endovascular techniques can rapidly control, improve or cure (with a reasonable chance of normal development) most of these lesions and eliminate their symptoms. Morbidity and mortality are very low if embolization is performed following proper indications and scheduled in accordance with a strict protocol (Fig. 3).

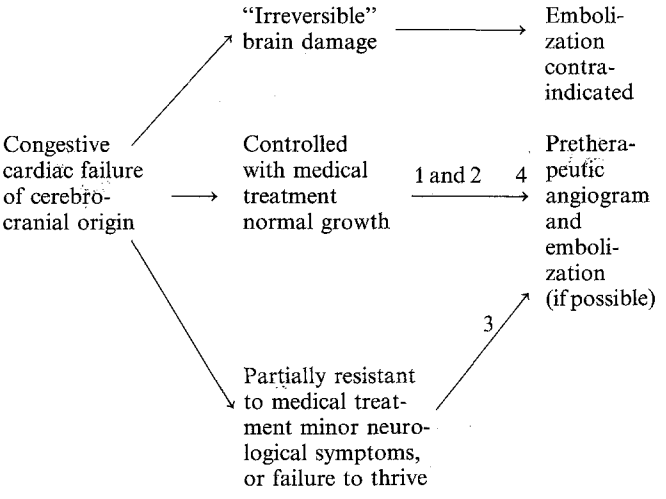

Fig. 3. Current protocol for the treatment of congestive cardiac failure of cerebrocranial origin. *1* Periodic clinical control, *2* periodic tomography and magnetic resonance imaging control, *3* urgent, *4* at 6–8 months

Patients are best managed in a pediatric intensive care unit. Pediatric neurosurgical and neurosurgical specialities should be readily available to support the interventional neuroradiology team.

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