Eur J Pediatr (1984) 142:298–300

Arteriovenous malformations as a cause of congestive heart failure in the newborn and infant

Three cases with different haemodynamic mechanisms

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Abstract. Three cases of congestive heart failure are presented caused by a cerebral arteriovenous aneurysm, a pulmonary arteriovenous aneurysm and a placental angioma respectively. Such conditions should be borne in mind in otherwise unexplained heart failure in the neonatal period and in infants. The haemodynamic influence of the malformation depends on its anatomy, giving rise to various clinical pictures.

Key words: Arteriovenous malformations – Congestive heart failure – Newborn

Introduction

Congestive heart failure in the neonatal period is usually caused by congenital heart disease. If congestive heart failure is not explained by a congenital cardiac defect or myocardial disease, the possibility of an extra-cardiac arteriovenous malformation should be taken into consideration [7]. In the following, three cases of abnormal congenital vascular channels are presented in which the clinical courses differed totally. This could be explained by the difference in haemodynamic mechanisms.

Case reports

Case 1. During the actual pregnancy the mother was treated with Thycapzol for hyperthyroidism. Birth was by caesarian section, birth weight was 3100 g. During the 1st day of life the girl became tachypnoeic and pale-greyish in colour and a rough systolic murmur was heard over the heart. The heart rate was 132 and respiration rate was 96. There was hepatomegaly (3 cm below the costal margin) and an X-ray of the chest showed cardiomegaly (cardiac/thoracic ratio 0.81) with normal pulmonary vasculature. The ECG showed a left-sided hypertrophy pattern for age. On the 4th day of life convulsions occurred. Because of intractable heart failure, congenital heart disease was suspected and heart catheterisation was carried out on the 5th day of life. Catheterisation data are shown in Table 1. Angiocardiography showed a large innominate artery with rapid opacification of the superior vena cava, which gave suspicion of an intra-cranial arteriovenous shunt. There were no signs of an inter-atrial shunt or other cardiac abnormalities. Further examination revealed a loud cranial bruit most clearly heard on the right side. A right-sided carotid arteriography showed a large aneurism of the vein of Galen (Fig. 1). A craniotomy was carried out and partial ligation of the vascular malformation was performed, but the infant died shortly after the operation from bradycardia and asystolia. Autopsy was not performed.

Case 2. The infant, a boy, was born at term with a birth weight of 1900 g after an uneventful pregnancy. The delivery and neonatal period were normal. At 2 months of age the infant was admitted to the paediatric department due to regurgitation of feeds and failure to thrive. The infant had normal colour but appeared hypertonic with hyperactive tendon reflexes. There was no cyanosis. The heart rate was 115 and respiration rate was 50. A rough systolic murmur was heard on the left side of the sternum and widespread over the back. An X-ray of the chest showed cardiomegaly (cardiac/thoracic ratio 0.55) and increased pulmonary vasculature. Congenital heart disease was suspected and heart catheterisation was carried out at 3 months of age. Catheterisation data appears in Table 1. No cardiac defect was found, but angiocardiography showed a pulmonary arteriovenous aneurysm in the upper lobe of the right lung (Fig. 2). The feeding difficulties gradually decreased and no specific treatment was instituted. The child developed mental deficiency and spastic tetraplegia. At 2 years of age a slight systolic murmur was still heard, but the cardiomegaly had disappeared. Telangiectasis on lips or in the nose or mouth was not observed.

Case 3. The birth was normal except for hydramnion 3000 ml. The birth weight was 3190 g. The placenta was abnormal with an angioma measuring $8 \times 5 \times 5$ cm (Fig. 3). The infant, a girl, was admitted to the paediatric department shortly after birth because of tachypnea and a short systolic murmur on the left side of the sternum. Thrombocytopenia (80×10^9/l)

Table 1. Catheterisation data in cases 1 and 2. The pulmonary arteries and left ventricles were not catheterised

	Case No. 1	5 days old	Case No. 2	101 days old
	Oxygen saturation per cent	Pressure mm Hg	Oxygen saturation per cent	Pressure mm Hg
Inferior vena cava	66		60	
Superior vena cava high	94			
Superior vena cava low	86		72	
Right atrium	90	9	68	7
Right ventricle apex	91	64/8	73	31/4
Right ventricle conus	92			
Left atrium		9	98	7

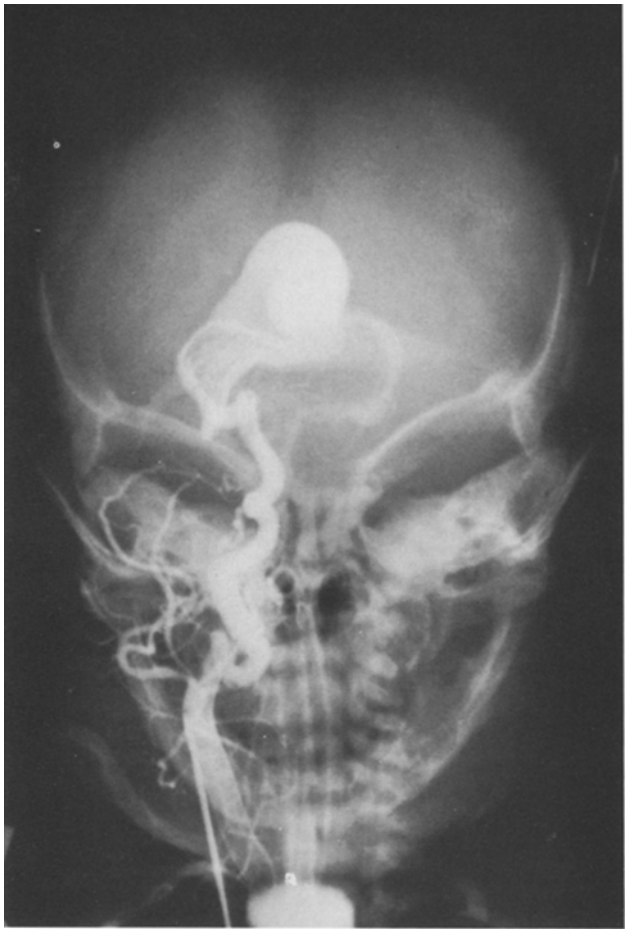

Fig. 1. Right-sided carotid arteriography in case No. 1 shows the arteriovenous malformation of the vein of Galen

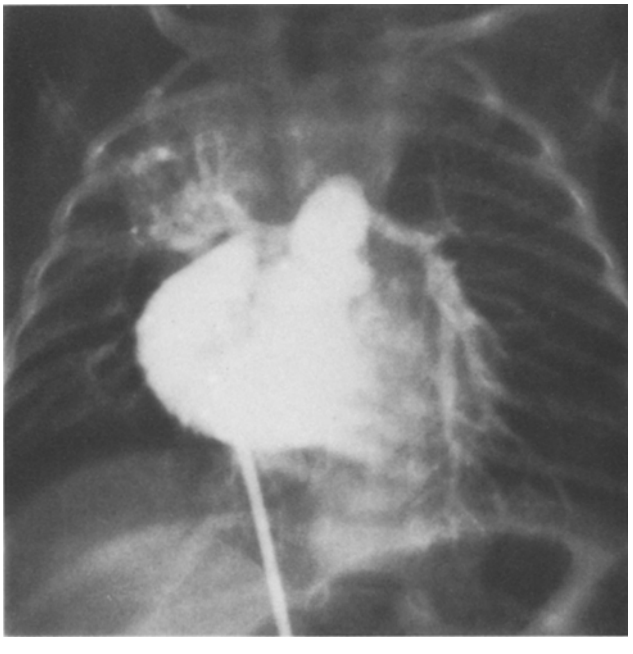

Fig. 2. Angiocardiography in case No. 2 shows the arteriovenous aneurysm in the upper lobe of the right lung. Injection was made in the right atrium

was present and there was hepatomegaly (5 cm below costal margin) and an X-ray of the chest showed cardiomegaly (cardiac/thoracic ratio 0.7) and hypervascularity of the lungs. An ECG showed a sinus rhythm of 140 per min, normal QRS axis in the frontal plane and no hypertrophy pattern. The infant was treated with digoxin and diuretics and improved quickly. The cardiomegaly and hypervascularity of the lungs decreased in a few days and the medical treatment was discontinued. Heart catheterisation was not carried out. At 3 years of age there was no detectable murmur and the X-ray of the chest was normal.

Discussion

Congestive heart failure in infants with extra-cardiac arteriovenous malformation is sometimes more severe and resistant to usual medical treatment than congestive heart failure associated with intra-cardiac anomalies, as myocardial damage often is present in the former group [5]. Furthermore, it is possible that the normal newborn has an inappropriate adjustment of cardiac output to changes in pre-load and after-load [3]. Cerebral arteriovenous aneurysms of the vein of Galen is a rare disease. During a 21 year period, only 21 cases causing heart failure were seen in one major centre of paediatric cardiology [7]. In intracranial arteriovenous fistulas, the lesion is located rostrally and the blood is shunted into the venous system and returned to the right heart and via the lungs to the left heart, imposing an increased volume load on both sides of the heart. The shunt may be large enough to cause an abnormal flow distribution in the fetus with retrograde flow in the aortic arch [2]. The intracranial blood flow may be ernormously elevated, as demonstrated in case No. 1, but a decrease in the effective cerebral perfusion and compromised autoregulation could be the cause of neurological symptoms. In addition the latter may be caused by the vascular tumour as a space-occupying lesion. Successful treatment leads to cessation of the cardiac symptoms [7, 8].

Congenital pulmonary aneurysms are also rare [6]. Some cases have teleangiectasis of the Osler-Weber-Rendu variety [10]. Heart failure has been described in cases with an aortic supply of part of the right lung [9]. However the shunt in most instances is directed from

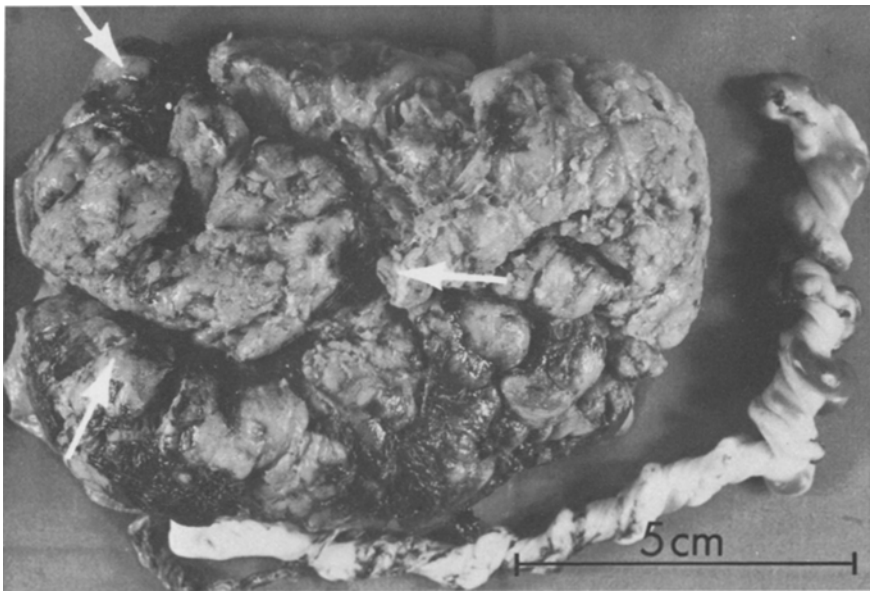

Fig. 3. Photo of the placenta from case No. 3. Arrows indicate the angioma

the pulmonary artery to the pulmonary vein, as in case No. 2, causing an increased volume load to the right heart. With the physiological, postnatal lowering of pulmonary vascular resistance the flow in the vascular malformation will gradually be reduced. This might explain why symptoms from the heart were only present during the first few months of life. At the age of 3 months physiological postnatal lowering of pulmonary vascular resistance is finished and heart catheterisation performed at that age thus only detected signs of right-sided heart failure while right to left shunting in the lungs of increased cardiac output was not prominent.

In the case of an arteriovenous shunt in the placenta the haemodynamic disturbance is only present before birth. A placental angioma as the cause of neonatal congestive heart failure is very rare [1, 11]. In most cases there is hydramnion [4]. Blood is shunted from the caudal aorta into the umbilical vein. From here it flows to the inferior vena cava, the right atrium, and through the foreamen ovale to the left atrium and left ventricle thus imposing an increased volume load on the left heart, which in prenatal life normally has a lower output than the right heart. This might damage the left heart predominantly, but as the caudal aorta is fed mainly from the right heart via the ductus arteriosus in the fetus, the volume load is on the heart as a whole. In case No. 3 symptoms appeared only in the first few days indicating that the haemodynamics normalised rapidly after birth. The only permanent change was a negative or diphasic T-wave in V_1-V_5 in the precordial leads of the ECG.

These cases demonstrate the old and wellknown fact that congestive heart failure in the neonatal period may exist without primary cardiac disease, but the clinical course can be quite diverse in different cases depending on the anatomy of the malformation and different haemodynamic mechanisms.

Acknowledgments. Johan Andersen MD, Institute of Pathology, Esbjerg County Hospital has kindly given permission to use the photograph in Fig. 3.

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Received January 9, 1984 /
Accepted April 9, 1984