A neonate with an unusual pulse discrepancy

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A 38-week gestational age boy was admitted after birth by Caesarean section to a neonatal unit. At 6 hours of life, a dextrose infusion was started because of hypoglycaemia. At 36 hours he developed cyanosis and tachypnoea. Physical examination revealed tachycardia, 2/6 systolic murmur, weakly palpable limb pulses, hepatomegaly and pulmonary rales. Laboratory tests were within the normal range, except blood gas analysis which showed respiratory and metabolic acidosis. Management entailed fluid resuscitation, mechanical ventilation and antibiotic administration. Because of recurrent metabolic acidosis and unstable haemodynamics, the infant was transferred at 48 hours to our neonatal intensive care unit with a diagnosis of probable sepsis. On admission all limb pulses were absent but carotid pulses were weakly palpable and became strong after establishing stable haemodynamics with inotropic support. Chest X-ray showed cardiomegaly and pulmonary oedema. Echocardiography confirmed the clinical diagnosis. The clinical condition of the patient improved quickly after initiation of treatment. Twenty-four hours later he was catheterized because of associated lesions. Left ventricular angiography is shown in figure 1. Retrograde aortography with a balloon-tipped catheter passed from the main pulmonary artery through the ductus arteriosus into the descending aorta (figure 2) confirmed the diagnosis. He was then prepared for surgery.

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Figure 1 Left ventricular angiography: lateral view

Figure 2 Retrograde aortography, frontal projection

Questions

- 1 What is your diagnosis ?
- 2 What do left ventricular angiography and retrograde aortography demonstrate ?
- 3 What is the present drug of choice for treatment ?

Answers

QUESTION 1

Physical findings of radial, carotid and femoral pulse discrepancy are typical of interrupted aortic arch or ductal-dependent coarctation. In this situation, where all limb pulses are very weak or impalpable and while both carotid pulses are strong, one would suspect interrupted aortic arch with obstruction proximal to the left subclavian artery with aberrant origin of the right subclavian artery from the descending aorta. Ductal-dependent coarctation with involvement of the left subclavian artery in the coarctation, its origin below the lesion, or a localised stenosis of the vessel's origin, associated with aberrant origin of the right subclavian artery, is the second possible diagnosis.

QUESTION 2

Left ventricular angiography (figure 1) confirms that both carotid arteries arise proximal to the interruption. The appearance of contrast in the descending aorta through an associated malalignment-type ventricular septal defect and the ductus arteriosus makes the site of interruption evident. A balloon-tipped catheter with proximal side holes is passed from the main pulmonary artery through the ductus arteriosus into the descending aorta and a retrograde aortography with inflated balloon shows that both subclavian arteries arise distal to the interruption (figure 2).

QUESTION 3

Standard therapy with prostaglandin E1 must be started immediately for induction and maintenance of ductal patency.

Discussion

Interrupted aortic arch is a rare congenital heart disease with a prevalence of 1% of critically ill cardiac infants. It is defined as a complete separation of ascending and descending aorta. It comprises several different anomalies that relate to the pattern of branching of the brachiocephalic arteries.¹ Patients typically present with acute cardiovascular collapse after spontaneous closure of the ductus arteriosus in

Learning points

- ductal-dependent left heart obstructive lesions (interrupted aortic arch and severe form of coarctation) are the main causes of cardiac failure in the neonate and the only congenital heart defects for which clinical examination with analysis of the pulse discrepancy suggests the exact diagnosis
- anomalous origin of the right subclavian artery from the aorta distal to the normally positioned left subclavian artery is one of the more frequent congenital anomalies. It is frequently associated with congenital heart disease
- when an aberrant right subclavian artery is associated with interruption between carotid and subclavian arteries, only the palpation of the carotid pulses is helpful to differentiate interrupted arch from critical aortic stenosis or sepsis

the first days of life. Interrupted aortic arch and severe forms of coarctation are the main causes of cardiac failure in the neonate and are often at the root of multiple organ failure which worsens the prognosis.²

Anomalous origin of the right subclavian artery from the aorta distal to the normally positioned left subclavian artery is one of the more frequent congenital anomalies. Zapata *et al*³ reported associations of aberrant right subclavian artery with aortic arch anomalies, and found this anomaly in 3% of cases of interrupted aortic arch and 1% of coarctation of the aorta.

Ductal-dependent left heart obstructive lesions are the only congenital heart defects for which clinical examination suggests the exact diagnosis with the consequence that initial management may be optimal. Physical findings of radial, carotid and femoral pulse discrepancy, depending on branching pattern, are typical of interrupted aortic arch or ductal-dependent coarctation (table).^{4,5} Differential cyanosis, although theoretically possible, is uncommon.⁵ Retro-oesophageal right subclavian artery is not uncommon and absence of right radial pulse does not exclude aortic arch anomalies. When an aberrant right subclavian artery is associated with interruption between carotid and subclavian arteries, only the palpation of the carotid pulses is helpful to differentiate interrupted arch from critical aortic stenosis or sepsis.⁴

In this case the diagnosis should have been suspected before clinical deterioration; quick transfer under prostaglandin E1 therapy would have avoided the development of acute cardiovascular collapse.

Final diagnosis

Interrupted aortic arch.

Keywords: congenital heart disease; neonate; aortic arch abnormality

Table Diagnosis according to the pulse discrepancy

Right arm pulses	Right carotid pulse	Left carotid pulse	Left arm pulses	Femoral pulses	Diagnosis
+	+	+	+	–	IAA type A or COA
+	+	+	–	–	IAA type B or COA
+	+	–	–	–	IAA type C
–	+	+	+	–	IAA type A+ARSA or COA+ARSA
–	+	+	–	–	IAA type B+ARSA or COA+ARSA
–	+	–	–	–	IAA type C+ARSA
–	–	–	–	–	sepsis or aortic stenosis or IAA or COA in agonal condition

Abbreviations: IAA = interrupted aortic arch, type A = interruption distal to the left subclavian artery, type B = interruption between the left common carotid artery and the left subclavian artery, type C = interruption between the innominate artery and the left common carotid artery; COA = coarctation of the aorta; ARSA = aberrant right subclavian artery; + = strong pulse, – = weak or impalpable pulse.

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Pericardial tamponade in a 65-year-old woman

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A 65-year-old woman with a one-month history of weakness and malaise was admitted to hospital because of chest pain, confusion, sweating and severe dyspnoea. Body temperature, blood pressure, pulse rate, and respiratory rate were 35.7°C, 80/60 mmHg, 62 beats/min and 38 breaths/min, respectively. Physical examination showed tachypnoea, jugular venous distension, pulsus paradoxus, kussmaul sign, hepatomegaly and diminished heart sounds. Chest X-ray disclosed a symmetrical globular enlargement of the heart (figure 1). Electrocardiogram revealed a reduction in amplitude of the QRS complex and sinus rhythm. Echocardiogram showed a massive pericardial effusion with right atrial and ventricular diastolic collapse. The diagnosis of pericardial tamponade was suggested, and emergency treatment was performed.

Haematological studies revealed a haemoglobin level of 11.6 g/dl, a leucocyte count of $11.8 \times 10^9/l$, and erythrocyte sedimentation rate of 52 mm/h. Serum biochemical profile was in normal range, and tests for viral, rheumatic and collagen disease were negative. Mantoux test was also negative. After stabilizing the patient's clinical condition, further studies were performed to establish the aetiology. Pericardial fluid examination was unremarkable except for the high protein content (4 g/dl), and microbiological cultures and cytologies were negative.

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Figure 1 Admission chest X-ray

Questions

- 1 What is the most probable cause of the cardiac tamponade in this patient?
- 2 What are the major causes of pericardial tamponade?
- 3 How should the patient be managed?