Unusual presentation of more common disease/injury

Primary retrieval of a shocked neonate with duct-dependent circulation to tertiary care by air ambulance

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Summary

In the UK the incidence of congenital heart disease is approximately 7 per 1000 live births. The case of a neonate with shock due to an undiagnosed heart defect who was resuscitated in a rural GP surgery and transferred directly to a paediatric centre with specialist tertiary services is described. This case emphasises the importance of the recognition of the sick infant and demonstrates what can be achieved within the current framework for delivery of care. Helicopter emergency medical teams have training in many advanced practical skills, diagnostic skills and clinical experience, which facilitate transfer of the patient to a place of definitive treatment, reducing morbidity and mortality.

BACKGROUND

The incidence of congenital heart disease varies between 6.9 and 8 per 1000 live births.¹ Not all variants are detected antenatally. Prehospital teams may encounter neonates with shock and cyanosis due to undiagnosed congenital heart disease. Such cases require prompt treatment in a specialist centre.

CASE PRESENTATION

A 10-day-old baby presented to a rural GP practice with poor feeding and respiratory distress. In view of the baby's condition the attending paramedic requested the Helicopter Emergency Medical Service (HEMS). The baby was cyanosed despite high flow oxygen (SpO₂ 84%) and was tachypnoeic (respiratory rate 70 breaths/min). Heart rate was 170 beats/min (sinus), capillary refill time prolonged (5 s) and there was extensive skin mottling. There was no murmur and blood sugar concentration was in the normal range.

TREATMENT

A fluid bolus was administered intravenously (20 ml/kg 0.9% saline) and capillary refill improved. Breathing was inadequate and the decision was taken to intubate and ventilate the infant.

Anaesthesia was induced with ketamine (2 mg/kg) followed by rocuronium (1 mg/kg). Post intubation and ventilation the baby remained cyanosed, SpO₂ 78% (FiO₂ 1.0), EtCO₂ 3.8 kPa with bilateral air entry on auscultation. Heart rate was 140 beats/min, systolic blood pressure 83 mm Hg and capillary refill 3 s. Primary transfer was made to Birmingham Children's Hospital (Birmingham, UK), which offers an emergency department (ED), paediatric intensive care unit (PICU) and cardiology. Full monitoring was continued, anaesthesia maintained with two ketamine boluses (1 mg/kg), and two further fluid boluses were given (each at 10 ml/kg) for prolonged capillary refill time.

On arrival, ED and PICU teams were consultant led. The patient remained cyanosed. A prostaglandin E2 infusion was given and transthoracic echocardiogram demonstrated transposition of the great arteries (TGA), with an intact ventricular septum. Despite blood flow through the patent ductus arteriosus the baby remained cyanosed. The systemic circulation was severely compromised and a significant lactic acidosis was demonstrated.

OUTCOME AND FOLLOW-UP

An urgent balloon atrial septostomy (4 mm) was performed through a tiny patent foramen ovale (PFO) under echocardiographic guidance. The clinical status subsequently improved rapidly, and the patient was discharged from the PICU to the ward the following day. After 5 days an 'arterial switch' operation was performed and baby made an uncomplicated recovery.

DISCUSSION

Transposition of the great arteries

TGA is a cyanotic variant of congenital heart disease occurring in 20–30 per 100 000 live births. The ventriculoarterial connections are discordant, resulting in the systemic and pulmonary circulations running in parallel, rather than in series (figure 1).² Survival is only possible if mixing occurs between the two circulations. Central cyanosis is observed clinically, with the onset and severity dependent on anatomical and functional variants that influence the degree of mixing between the two circulations. In this case, the circulation was maintained for the first few days of life by virtue of mixing via the small atrial septal defect. Once the ductus arteriosus started to close, the mixing at atrial level reduced, resulting in severe hypoxaemia. Oxygen delivery was therefore impaired, leading to shock.

Treatment with intravenous prostaglandin E1 or E2 maintains ductal patency promoting intercirculatory mixing via the PFO. However, if the PFO is restrictive (too

What is transposition of the great arteries?

The normal heart

Transposition of the great arteries

Transposition of the great arteries – or TGA for short – means that the pulmonary artery arises from the left ventricle instead of the right, and the aorta arises from the right ventricle instead of the left. See the illustrations above.

Figure 1 Transposition of the great arteries (reproduced with the kind permission of the British Heart Foundation, the copyright owner).²

small), mixing will be inadequate. Urgent transfer to a paediatric cardiac centre is necessary and usually performed by a designated neonatal or paediatric retrieval service.

Balloon atrial septostomy (Rashkind procedure) is an emergency procedure to increase the interatrial communication. The procedure involves positioning a balloon-tipped catheter into the left atrium via the PFO. The balloon is then inflated and pulled back into the right atrium, tearing the atrial septum, allowing intracardiac mixing of pulmonary and systemic blood. This procedure allows stabilisation of patients prior to corrective surgery (arterial switch operation).

The prognosis of TGA without surgery is poor (1 year survival <10%). Corrective surgery is performed to achieve physiological and anatomical repair. Long-term survival is very encouraging (>90% at 15 years) and the reintervention rate is low.³ Long-term follow-up is required.

Prehospital retrieval

'Medical cases' form a significant workload for the HEMS, especially in rural areas. To be called to a shocked duct-dependent (undiagnosed) neonate in the prehospital environment is a rare occurrence. The key to survival and successful management is the initial recognition of the sick infant. Transportation options need to be timely and initial resuscitation may be complex and skill dependent.

Early identification of the need for critical care for this baby enabled critical interventions at scene and definitive care to be sought by HEMS personnel. The direct transfer of the baby to a tertiary centre removed the need for secondary transfer by the neonatal retrieval service (hospital based) in what proved to be a time-critical condition.

Learning points

- This case emphasises the importance of the recognition of the sick infant and demonstrates what can be achieved within the current framework for delivery of care.
- Helicopter Emergency Medical Service (HEMS) teams (doctors and paramedics) have training in many advanced practical skills enhancing delivery of critical care interventions.⁴
- Diagnostic skills and clinical experience should facilitate transfer of the patient to a place of definitive treatment, reducing morbidity and mortality.

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Competing interests None.

Patient consent Obtained.

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