CASE REPORT

Persistent pulmonary hypertension of the newborn associated with pulmonary atresia and intact interventricular septum

M Codispoti, J E Burns, S G Haworth, D Simpson, P S Mankad

Abstract

Neonates with pulmonary atresia and intact interventricular septum (PAIVS) do not have pulmonary vascular disease secondary to their heart abnormality. Persistent pulmonary hypertension of the newborn has not been described in association with this condition. The case is reported of a female neonate born with PAIVS, who preoperatively had no clinical evidence or any risk factors for persistent pulmonary hypertension of the newborn, but whose postoperative course was highly suggestive of persistent pulmonary hypertension; necropsy confirmed the features of pulmonary vascular disease.

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Keywords: persistent pulmonary hypertension; pulmonary atresia and intact ventricular septum; pulmonary vascular disease; surgery; congenital heart defects

Department of Cardiac Surgery, Royal Hospital for Sick Children, Sciennes Road, Edinburgh EH9 1LF, UK
M Codispoti
J E Burns
D Simpson
P S Mankad

Division of Developmental Cardiology, Institute of Child Health, Hospital for Sick Children, Great Ormond Street, London WC1N 3JH, UK
S G Haworth

Correspondence to:
Mr Mankad.
email: pankaj.mankad@ed.ac.uk

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Neonates with pulmonary atresia and intact interventricular septum (PAIVS) do not have pulmonary vascular disease secondary to their heart abnormality.[1] Failure of the pulmonary vasculature to adapt to extrauterine life, giving rise to persistent pulmonary hypertension of the newborn, has not been described in association with PAIVS.

Case report

A newborn female was transferred to our hospital with a suspected cyanotic congenital heart defect. The pregnancy, labour, and delivery were all normal. The baby was born at term weighing 3240 g, and no active resuscitation was needed. Apgar score was 8 at one minute and 9 at five minutes. There was no hypothermia, hypocalcaemia, hypoglycaemia, or polycythaemia, and she did not appear postmature. The mother's health had been excellent, with no gestational diabetes or history of drug intake, including anti-inflammatory agents and analgesics.

Echocardiography confirmed valvar pulmonary atresia and intact interventricular septum. The right ventricle was tripartite and reasonable in size. The tricuspid valve annulus was 5 mm in diameter, just 1 SD below the normal mean value for her weight (Z value –1). This was considered adequate for antegrade decompression of the right ventricle, although the tricuspid inflow could not be assessed owing to absence of regurgitation and lack of pulmonary outflow. The infant was stabilised with prostaglandin E_2 infusion (10 ng/kg/min), maintaining arterial oxygen saturations between 80% and 90% on room air in the preoperative period. At age 4 days, the baby underwent transpulmonary pulmonary valvectomy with autologous pericardial patch closure of the pulmonary artery on cardiopulmonary bypass and induced ventricular fibrillation. At operation the pulmonary valve was found to be very dysplastic and unicuspid. Pulmonary valvotomy would have left significant residual obstruction and pulmonary incompetence because of the unicuspid nature of the valve; therefore, this was excised. There was some anterior deviation of the outlet septum, but this was not causing any subvalvar obstruction. Following valvectomy, the pulmonary artery annulus admitted a 6 mm Hegar dilator. During surgery the aorta was not cross clamped and the ductal flow was controlled temporarily with a snare. Cardiopulmonary bypass was discontinued uneventfully.

The infant's initial postoperative course was stable, although arterial oxygen saturation was persistently lower than before surgery. This was considered acceptable following cardiopulmonary bypass, and the infusion of prostaglandin E_2 was continued to maintain ductal patency, with the plan that it be weaned after a few days, once right ventricular function had improved following adequate relief of outflow obstruction. She remained on 3 μg/kg/min dopamine and 4 μg/kg/min glyceryl trinitrate infusion. During mechanical ventilation with FiO_2 between 60% and 80%, her arterial oxygen saturation was stable, averaging 70% (rarely above 75%) until the third postoperative day when there was a further deterioration: arterial oxygen saturation was close to 60% and dipped to the high 50s, associated with sinus tachycardia. Echocardiography showed that the duct was widely patent, but flow velocities were low and most of the ductal flow was retrograde into the right ventricle. It

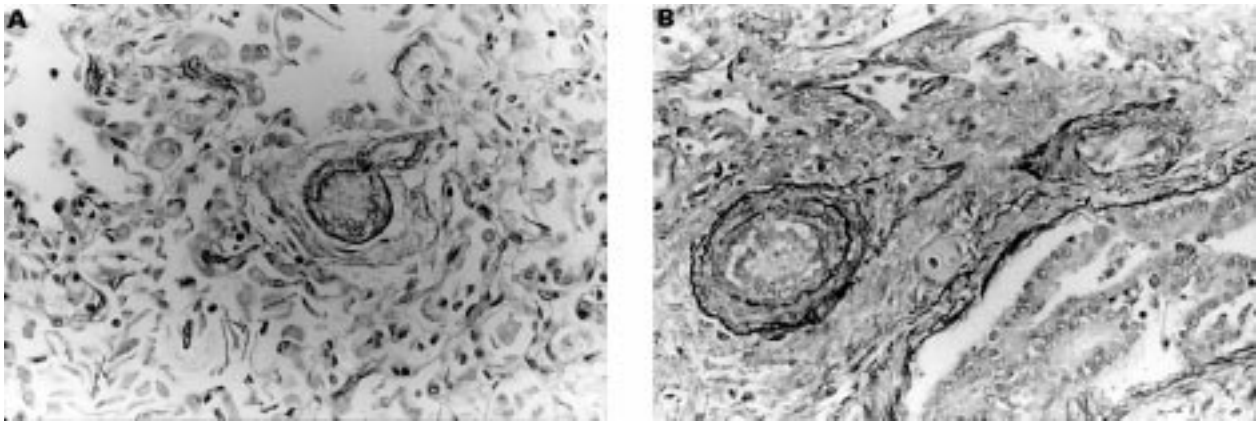

Figure 1 (A) Photomicrograph of lung tissue showing an alveolar duct artery with an abnormal thick media and muscle extending into a small precapillary branch. (Magnification: 4 mm = 10 μm.) (B) Terminal bronchiolar artery, on the left, with medial hypertrophy and intimal proliferation. Its branch, on the right, also showed medial hypertrophy. (Magnification: 4 mm = 10 μm.)

appeared that pulmonary vascular resistance was raised causing the ductal flow to enter the low resistance right ventricle. Inhaled nitric oxide (20 ppm), was therefore started, which resulted in an immediate dramatic improvement in arterial oxygen saturation from 50% to 80% on the same inspired oxygen concentration. Even though arterial pressure was lower than at the time of the previous echocardiogram, the Doppler velocity across the duct increased from 2 m/s to nearly 3 m/s with nitric oxide. Inhaled nitric oxide was maintained throughout the postoperative period, with minor adjustments in inotropic support. Twenty four hours later, the dependency of arterial oxygen saturation on inhaled nitric oxide was once again confirmed, both clinically and by ultrasound examination. Echocardiography suggested that the pulmonary artery pressure was still high, despite inhaled nitric oxide, and that both ventricles were underfilled, although with excellent systolic function. A period of relative stability followed until the seventh postoperative day when she suffered a sudden clinical deterioration, which resembled a pulmonary hypertensive crisis (arterial pressure 60/35 mm Hg, central venous pressure 14 mm Hg, oxygen saturation 55%) from which she could not be resuscitated.

Necropsy showed satisfactory pulmonary valvectomy, with the annulus patent to an 8 mm Hegar dilator, and good sized main, right, and left pulmonary arteries (5 mm in diameter). The ductus arteriosus was patent and the lumen was at least 3 mm in diameter. The tricuspid valve leaflets were thickened and partly fused, with thick and short chordae tendinae. The histology of the right ventricle was consistent with outflow obstruction. However, the most interesting features were seen in the pulmonary vasculature. Neonates with pulmonary atresia are usually born with abnormally thin walled pulmonary arteries,[1] but, in this baby, the pulmonary vessels were abnormally thick walled and in many vessels the muscular hypertrophy extended into smaller, more peripheral arteries than normal (fig 1A). Early intimal thickening was seen in several terminal and respiratory bronchiolar arteries (fig 1B). These findings were present in both lungs but were much more severe in the left lung. The lymphatic channels in the right lung were hugely distended both in the alveolar septa and in the alveolar walls. These necropsy findings in the pulmonary vasculature were unusual, and the observed structural abnormalities in the lungs were in accordance with her clinical picture.

Discussion

There have been significant advances in our understanding of the morphology and physiology of neonates with PAIVS. This has contributed to a logical approach to the initial management of these babies by catheter intervention, pulmonary valvotomy, transannular patching, with or without systemic to pulmonary artery shunt.[2 3] Despite this differential approach, significant mortality still follows the initial management of these babies.[4] This often results from right ventricular dysfunction or persistent arterial desaturation, in the absence of an adequate source of pulmonary blood flow.

In our patient, the pulmonary outflow tract was enlarged and repaired to achieve a diameter that would have allowed sufficient flow to the pulmonary circulation, if this had been normal. At necropsy, there is rarely a compelling reason to examine pulmonary parenchyma in detail, as PAIVS is not associated with abnormalities in the pulmonary vasculature. However, in this infant the clinical picture prompted us to perform a detailed pathological examination of the lungs. The microscopic observation of the lung parenchyma uncovered the rare association of pulmonary arterial medial hypertrophy with PAIVS. We speculate that this mechanism might also account for our observation that occasional neonates operated for duct dependent pulmonary atresia/severe tetralogy of Fallot seem to need a very high systemic arterial pressure (100–120 mm Hg systolic) in the early postoperative period to maintain adequate oxygenation. It is conceivable that some of these babies have associated persistent

pulmonary hypertension of the newborn. We therefore feel that this association should be considered if the early postoperative course suggests this possibility in any infant, even with reduced pulmonary blood flow.

1 Haworth SG, Reid L. Quantitative structural study of pulmonary circulation in the newborn with pulmonary atresia. *Thorax* 1977;**32**:129–133.
2 Just RN, Nykanen DG, Williams WG, *et al*. Transcatheter perforation of the right ventricular outflow tract as initial therapy for pulmonary valve atresia and intact ventricular septum in the newborn. *Cath Cardiovasc Diagn* 1997;**40**: 414–18.
3 Pawade A, Karl T. Management strategy in neonates presenting with pulmonary atresia and intact ventricular septum. *Curr Opin Cardiol* 1994;**6**:600–5.
4 Bull C, Kastelka M, Sorensen K, *et al*. Outcome measures for the neonatal management of pulmonary atresia with intact ventricular septum. *J Thorac Cardiovasc Surg* 1994;**107**:359–66.

Electronic Pages

eHEART: *www.heartjnl.com*

The following electronic only articles are published in conjunction with this issue of *Heart*.

Heart transplant for dilated cardiomyopathy associated with polymyositis

A Afzal, R S D Higgins, E F Philbin

Cardiac involvement is one of the most significant factors in the poor clinical outcome of polymyositis. The case of a 39 year old African American woman with polymyositis, cardiomyopathy, and severe heart failure who had orthotopic heart transplantation is described. Review of the literature reveals that cardiac manifestations of polymyositis are frequent and include conduction system abnormalities, myocarditis, cardiomyopathy, coronary artery atherosclerosis, valvar disease, and pericardial abnormalities. (*Heart* 1999;**82**:e4) www.heartjnl.com/cgi/content/full/82/4/e4

Non-coronary sinus of Valsalva aneurysm diagnosed after a road traffic accident

Â Ribeiro, F Fernandes, A Costeira, A Simões, P Rodrigues

A 38 year old man with a huge unruptured sinus of Valsalva aneurysm, complicated with severe valvar aortic regurgitation, is described. The aneurysm was detected by echocardiography in the asymptomatic patient who presented with an intense precordial diastolic rumble after a road traffic accident. The patient had successful surgery for the aneurysm and aortic valve replacement. Possible aetiologies for the aneurysm and a brief revision of clinical aspects and treatment are discussed. (*Heart* 1999;**82**:e5) www.heartjnl.com/cgi/content/full/82/4/e5

Salvage angioplasty and stenting following spontaneous dissection of the left anterior descending coronary artery

A P Banning

A 46 year old woman presented with sudden onset severe, tight, central chest pain. ECG showed sinus rhythm with marked ST elevation over the anterior chest leads, and she received intravenous tPA within two hours of symptom onset. The pain persisted despite thrombolysis, diamorphine, and intravenous β blockade. Coronary angiography showed an occlusive dissection flap in the proximal left anterior descending coronary artery. Balloon angioplasty resulted in improved flow and two overlapping intracoronary stents were placed in the proximal and mid-vessel. When the procedure had been completed it became apparent that the dissection had involved the entire vessel up to the apex of the left ventricle. An intra-aortic balloon pump was inserted for 48 hours and the patient was subsequently discharged with treatment of an angiotensin converting enzyme inhibitor, β blocker, aspirin, and ticlopidine. (*Heart* 1999;**82**:e6) www.heartjnl.com/cgi/content/full/82/4/e6