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Brief Report

Neonatal death due to transposition in association with premature closure of the oval foramen

Michael Ashworth, Mudher Al Adnani, Neil J. Sebire

Department of Paediatric Histopathology, Great Ormond Street Hospital, London, United Kingdom

Abstract We present a male infant, born at term, who collapsed and died at 30 minutes of age. Autopsy demonstrated concordant atrioventricular and discordant ventriculo-arterial connections with premature closure of the oval foramen, which bulged markedly into the left atrium. Premature closure of the oval foramen is rare in the setting of transposition. As far as we know, there have been only three previously reported cases.

Keywords: Congenital heart disease; cyanosis; neonatal collapse

NEONATAL COLLAPSE WITH CYANOSIS IS A recognized presentation of previously unrecognized congenital cardiac disease, of which transposition is one subtype. In most such cases, the initially patent oval foramen and arterial duct allow sufficient haemodynamic compensation for stabilisation to be achieved prior to surgical repair. We present a patient who died in the neonatal period with transposition. The patient had suffered premature closure of the oval foramen. We discuss the pathology of this association of anomalies.

Case report

A male infant was born at 39 weeks gestation by spontaneous vaginal delivery, following an uncomplicated antenatal course, and with an apparently unremarkable ultrasonic examination carried out in the second trimester at the local hospital. There was no significant previous obstetric or medical history, and labour was reported as uneventful, with APGAR scores being nine at one minute, and 10 at five minutes. The baby weighed 3.2 kilograms at birth. He was well until around 30 minutes of age, when increasing grunting was noted. On examination at this time, he was noted to be markedly cyanotic and bradycardic, and did not respond to basic life-support measures. He was intubated, and further resuscitation was attempted with adrenalin and surfactant. Blood gases measured in a sample taken from the umbilical vein demonstrated acidosis, with pH of 6.8. His saturations of oxygen remained poor, and he was commenced on high-frequency oscillatory ventilation and 100% oxygen. Despite these measures, he remained severely bradycardic and cyanotic with worsening acidosis, continued rapidly to deteriorate, and died.

At autopsy, there were no significant abnormalities on external examination. Internally, the heart showed usual arrangement of the atrial appendages, with the apex directed inferiorly. The systemic and pulmonary venous connections were normal. The atrioventricular connections were concordant, but the ventriculo-arterial connections were discordant, with the aorta lying anteriorly and to the right of the pulmonary trunk. The aortic arch was to the left, with a normal pattern of branching. The arterial duct was closed, although it still admitted a thin probe. The oval foramen was closed, the flap valve being sealed along its edges, with the floor of the fossa bulging into the cavity of the left atrium. The right ventricular wall was hypertrophied and the right ventricle larger than the left. The atrioventricular and arterial valves were normal, and the ventricular septum was intact (Fig. 1).

Correspondence to: Michael Ashworth, Department of Paediatric Histopathology, Great Ormond Street Hospital, Great Ormond Street, London WC1N 3JH, United Kingdom. Tel: +0207 829 8663; Fax: +0207 829 7875; E-mail: AshwoM@gosh.nhs.uk

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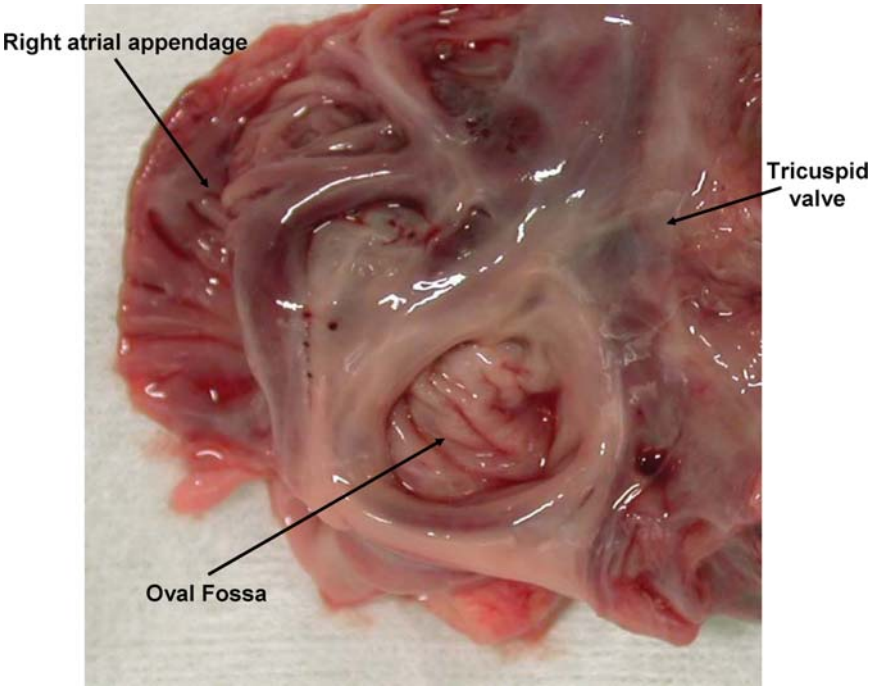

Figure 1.
Right atrium viewed from the right side. The oval fossa shows a wrinkled and collapsed closed flap valve.

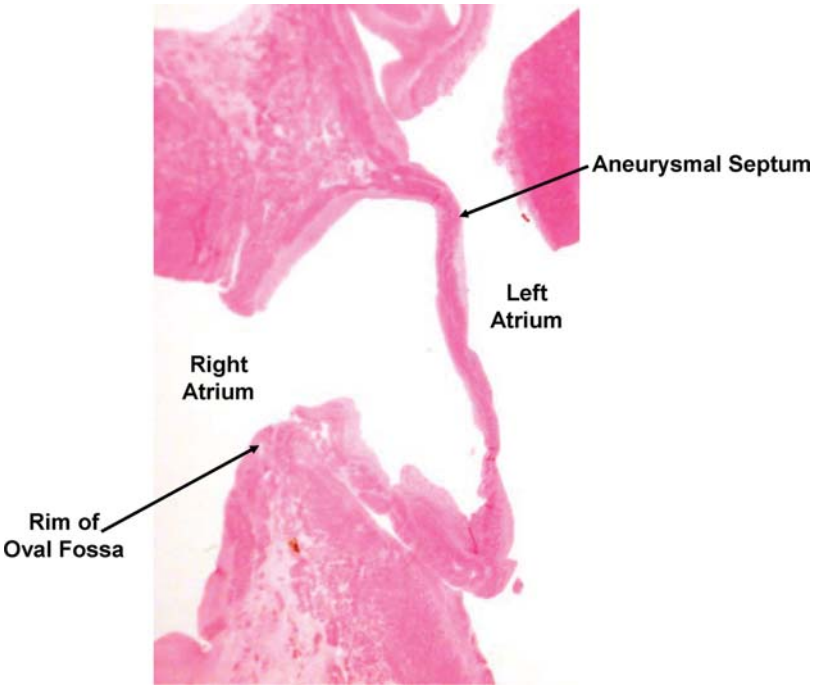

Figure 2.
Low power photomicrograph of a histological section through the interatrial septum showing the intact oval fossa and the aneurysmal bulge of the flap valve to the left.

Histologically, the right ventricular myocardium was hypertrophied, albeit without myofiber disarray. There was no necrosis, but there was intense capillary engorgement and focal interstitial haemorrhage. Histology confirmed the integrity of the oval fossa, with the floor bulging markedly to the left (Fig. 2). The lungs contained thick-walled peripheral muscular arteries, and there was interstitial haemorrhage. The features were those of transposition with an intact ventricular septum, along with premature closure of the oval foramen.

Discussion

Our patient suffered an uncommon cause of early neonatal death due to a combination of congenital

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cardiac defects. The oval foramen forms a vital part of the fetal circulation, permitting bypass of the lungs by oxygenated blood from the umbilical vein, with delivery to the left heart, and thence to the coronary arteries and developing brain. At birth, the foramen closes following full incorporation of the lungs into the circulation. Premature closure is rare, albeit responsible on occasion for fetal hydrops, and sometimes even intrauterine fetal death. In such cases, there is frequently hypoplasia of the components of the left heart, although it remains unclear as to whether this is a consequence or a cause of the premature closure. The mechanisms of premature closure are unknown.

Transposition, representing the combination of concordant atrioventricular and discordant ventriculo-arterial connections, is a serious malformation that requires the presence of a functioning postnatal right-to-left shunt to permit mixing of blood from the otherwise self-contained pulmonary and systemic circulations, this being needed to provide adequate delivery of oxygen. Such a shunt is usually provided by the initially patent oval foramen and arterial duct. With closure of these structures after birth, and in the absence of a ventricular septal defect, the circulation is unsustainable. Intervention, such as atrial septostomy, must then be performed to ensure survival before definitive surgical correction can be performed. Self-evidently, premature closure of the oval foramen will produce a circulation entirely dependent on the arterial duct for mixing of right and left-sided blood.

In the setting of transposition, therefore, closure of the oval foramen means that the entire venous return crosses the tricuspid valve, traverses the right ventricle, and reaches the aorta. The arterial duct supplies blood to the lungs, which because of the high pulmonary vascular resistance will receive only a small amount of flow. There is consequently reduced venous return to the left atrium, and hence a small left atrium and left ventricle. During fetal life, therefore, the right atrium and ventricle will usually be dilated because of increased volume load, with hypertrophy of the right ventricular myocardium. While this situation may be sustainable during fetal life, postnatally the circulation will become critically dependent on the patency of the arterial duct. With its usual neonatal closure, this conduit is functionally removed.

To the best of our knowledge, there are only three previously reported cases of premature closure of the oval foramen in the setting of transposition. Bhatt & Jue[1] reported the first case in 1979, noting the presence of normal left-sided cardiac components. Berry et al.[2] then reported a male infant, born at 36 weeks gestation, who developed cyanosis at birth, and suffered bradycardia and cardiac arrest shortly thereafter. At necropsy, the right ventricle was hypertrophied, the oval fossa was closed, and bulged to the left. Discordant ventriculo-arterial connections were confirmed. The left ventricle was small and the arterial duct hypoplastic. The most recent case was reported by Donofrio.[3] This baby was delivered by emergency caesarean section at 34 weeks gestation, subsequent to the routine antenatal scan at 20 weeks being reported as normal. At 34 weeks gestation, the mother presented with shortness of breath. Ultrasonic examination then showed transposition. The mitral valve was thickened and severely regurgitant, the atrial septum was intact and bowed to the right. No flow was seen in the arterial duct. There was minimal fetal movement and, because of concerns for fetal wellbeing, emergency caesarean section was undertaken. On delivery, the baby was limp and cyanotic and resuscitation was immediately commenced and the infant placed on extracoporeal membrane oxygenation at age six minutes. At seven days of age an arterial switch operation was performed. Within an hour of the operation, pulmonary hypertension developed and extracoporeal membrane oxygenation was recommenced, but the baby developed an intraventricular cranial haemorrhage and died.

Our patient demonstrates similar anatomical features, with an atrial septum bulging to the left, a closed arterial duct, hypertrophy of the right heart, and transposition. While it is theoretically possible to correct such a condition, there are at present no known survivors. The most recent report[3] does raise the possibility that, provided the condition is recognized antenatally, it may be possible to achieve a satisfactory outcome. Our case also demonstrates the problems in making the correct diagnosis with ultrasound, so that the condition may not come to medical attention until the fetus or mother is symptomatic, by which time it may well be too late to achieve an optimal outcome. Alternatively, presentation may be with sudden collapse on the first day of life, a clinical presentation which is not infrequently due to underlying unrecognized congenital cardiac disease. Transposition is well-recognized as such a cause, with the clinical presentation worsened by the premature closure of the oval foramen.

Although death during infancy due to congenital cardiac disease is well-described,[4] we have presented a rare case of unexpected early neonatal death due to the combination of transposition and premature closure of the oval foramen, leading to severe haemodynamic compromise. Our experience highlights the relatively common presentation of unsuspected congenital cardiac disease with neonatal collapse, and reiterates the importance of thorough post-mortem examination in such cases.

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