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Brief Report

Cardiac magnetic resonance imaging in a premature baby with interrupted aortic arch and aortopulmonary window

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Abstract Aortopulmonary window is a communication between the main pulmonary artery and the ascending aorta in the presence of two separate semilunar valves. The combination of an aortopulmonary window with interrupted aortic arch is rare. We discuss the unique case of an extremely premature infant weighing 1.7 kilograms who underwent cardiovascular magnetic resonance imaging as a pre-operative assessment in a high-field open 1.0 Tesla magnetic resonance imaging system as a one-stop investigation before complete repair.

Keywords: Congenital heart disease; interrupted aortic arch; aortopulmonary window; 1.0 Tesla magnetic resonance imaging system; cardiac magnetic resonance imaging

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Aortopulmonary window is a rare congenital heart disease entity. Several different classification schemes have been proposed, with the types of proximal, distal, total, and intermediate having been recommended most recently.[1] The combination of an aortopulmonary window with aortic arch interruption is also rare. It has been found that in patients with aortopulmonary window the coexistence of aortic arch interruption occurs in 13% of cases.[2] In addition, a congenital cardiac surgeons' society study has found that among 472 neonates with interrupted aortic arch, only 20 patients had an aortopulmonary window in association.[3]

The outcome of such complex congenital heart disease in very premature and low birth weight babies is usually guarded, and non-invasive cardiac imaging, wherever possible, is preferred over cardiac catheterisation.[4] Non-invasive imaging in the form of cardiac magnetic resonance imaging has been performed predominantly in closed-bore 1.5 Tesla systems, although not so frequently in very small and low gestational age infants.

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Clinical summary

A female baby, one of twins, was born prematurely by spontaneous delivery at 33 weeks of gestation. The baby weighed 1.7 kilograms and required no resuscitation at birth. She had only mild tachypnoea with no cyanosis and no oxygen requirement. Her blood pressure was 83/52 mmHg with no difference between extremities, respiratory rate 50–60 per minute, and heart rate 145–170 beats per minute. A grade 2–3/6 systolic murmur heard over the upper left sternal border raised the suspicion of cardiac disease. Transthoracic echocardiography showed mild dilation of the left heart cavities with preserved systolic function, mild mitral insufficiency, a secundum atrial septal defect of 5 mm, and no ventricular septal defect. An aortopulmonary window forming a large aortic sac was seen from the high parasternal long-axis view. The aortic arch also appeared interrupted, with two vessels seen before the interruption. A large ductus of the size of the pulmonary artery with bidirectional flow connected the pulmonary artery to

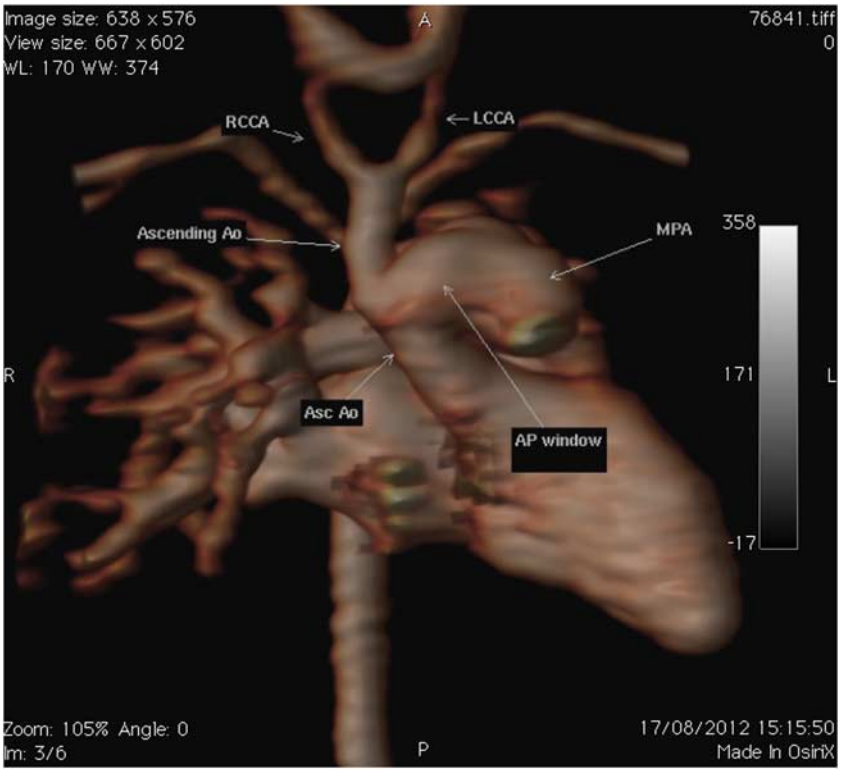

Figure 1.
Anterior view of the ascending aorta with the origin of the two carotids before the interruption and the aortopulmonary window connecting the main pulmonary artery with the ascending aorta. LCCA = left comman carotid artery; MPA = main pulmonary artery; RCCA = right common carotid artery.

the distal aorta from which the left subclavian artery arose. Another vessel also arose from the distal aorta, but its course could not be delineated. She was commenced on diuretics and captopril, and feeds were fortified in an attempt to increase her weight, before reparative surgery. However, tachypnoea and poor weight gain persisted, and hence it was considered that early surgery could not be avoided.

Further delineation of the anatomy was considered necessary before surgical repair. Owing to the neonate's low weight, an attempt with a non-invasive examination, such as cardiac magnetic resonance imaging, was considered more appropriate before embarking on cardiac catheterisation. Our patient was intubated and prepared for magnetic resonance imaging in the neonatal unit and was transferred to the magnetic resonance imaging department for a targeted examination. In our institution, we use a 1 Tesla open-magnet magnetic resonance imaging system (Panorama, Philips, The Netherlands). The patient underwent first-pass three-dimensional magnetic resonance angiography, as well as imaging with T1-weighted black-blood sequences in order to delineate the vascular relationships to the trachea and the oesophagus. For the magnetic resonance imaging angiography, 0.4 ml of Omniscan was used and the sequence was planned with field of view (FOV) = 160 mm, repetition time (TR) = 3.8 ms, echo time (TE) = 1.5 ms, and slice thickness = 3.0/−0.5 mm. The black-blood sequence was planned with TR = 400 ms, TE = 40 ms, slice thickness = 2.5/−0.5 mm, and number of signal averages (NSA) = 4. An aortopulmonary window was clearly demonstrated both on the three-dimensional angiogram (Fig 1) and on the black-blood sequences of the total type.[1] The right subclavian artery appeared to originate from a common vascular trunk arising from the posterior aspect of the ductal arch and travelling aberrantly to the right (Fig 2). Its course did not appear to cause any compression to the oesophagus or the trachea. The neonate was scheduled for surgery and was operated successfully 10 days after the magnetic resonance imaging examination. Cardiac catheterisation was not required before surgery.

Discussion

Although cardiac magnetic resonance imaging is currently the investigation of choice for delineation of complex anatomy in patients with congenital heart disease, it has not been used frequently in

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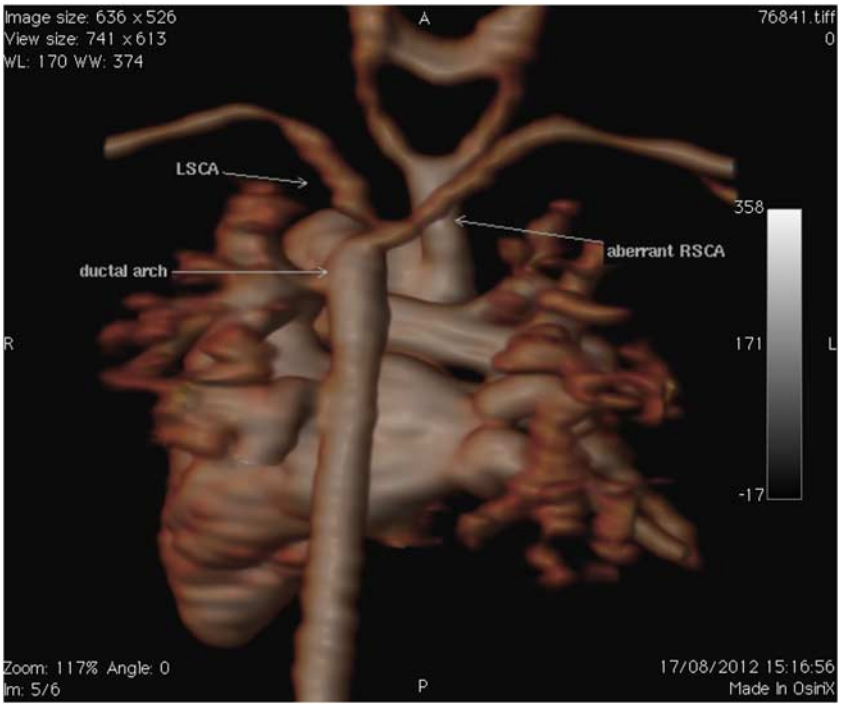

Figure 2.
Posterior view of the ductal arch and the common arterial trunk that gives rise to the left subclavian artery (LSCA) and the aberrant right subclavian artery (RSCA).

preterm babies with very low birth weight. This is due to the risk of hypothermia during the magnetic resonance imaging examination, but also the drawbacks of the magnetic resonance imaging environment, particularly of a closed-magnet system, which may potentially be unsafe because of the restricted access to the intubated patient.[5] Advanced cardiac magnetic resonance imaging is traditionally performed in closed-bore magnetic resonance imaging scanners of 1.5 Tesla, as it provides higher signal-to-noise ratio, which is proportional to the static magnetic field strength. All of the studies performed in very small infants that have been reported to date have taken place on a 1.5 Tesla scanner.[6,7] To the best of our knowledge, this is the first time that such complex anatomy is demonstrated with excellent image resolution in a high-field open 1.0 Tesla magnetic resonance imaging system, despite the extremely low weight of the neonate. This system has proved in our institution to be very good in imaging structural heart disease, where cine images, three-dimensional steady-state free precession, black-blood sequences, and three-dimensional magnetic resonance angiography are most commonly employed. In addition, it provides better access and visual, as well as mechanical, monitoring of the patient. This is particularly important in the case of intubated low-weight or gestational age infants.

Conclusions

Cardiac magnetic resonance imaging can be a one-stop investigation for children and infants with complex congenital heart disease and can be safely performed even in preterm small neonates under 2 kilograms. Although small infants have been previously scanned in a 1.5 Tesla closed-bore magnetic resonance imaging system, the high-field open 1.0 Tesla magnetic resonance imaging equivalent can also produce high-definition imaging. To this end, the open magnetic resonance imaging system might become an attractive alternative for some patients with structural heart disease because of the good image resolution and the unrestricted access provided to intubated infants and children.

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Supplementary material

For supplementary material referred to in this article, please visit http://dx.doi.org/doi:10.1017/S1047951112001461

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