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Case Report

# Complementary Use of Imaging modalities in Diagnosis of Complex Congenital Heart Disease

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## INTRODUCTION

Congenital heart disease (CHD) affects almost one in 100 newborn babies worldwide (Hoffman). The continuous advances and availability of non-invasive cardiac imaging has revolutionised the management of these cases, allowing various methods to complement one another. To have a better understanding of the varied and often-complex cardiac and extra-cardiac anatomy in patients with CHD, additional imaging may be required as a supplement to more traditional first-line modalities such as echocardiography (ECHO). In this report we present a case of an infant with a diagnosis of DiGeorge Syndrome, and tetralogy of fallot (TOF) with severe pulmonary stenosis and complex major aorto-pulmonary collateral arteries (MAPCAs). We will also outline the various roles of imaging modalities used in the management of this child, focusing on the complementary role of Multi-Detector Computed Tomography (MDCT).

## Case Report

A 5-month-old female with DiGeorge Syndrome, and TOF with near pulmonary atresia, presented with repeated episodes of collapse and severe respiratory distress. She was born at term via a non-complicated spontaneous vaginal delivery. The diagnosis of TOF was made antenatally and karyotyping confirmed a 22q11 deletion (DiGeorge) genetic abnormality.

On admission to our institution, she had symptoms of respiratory distress. Her parents reported a two-day history of coryzal symptoms. On examination she was cyanosed, with an arterial oxygen saturation of 50%. She was transferred to the Pediatric Intensive Care Unit (PICU). Chest x-ray showed right upper lobe consolidation, consistent with a lobar pneumonia, and antibiotic therapy was subsequently commenced. The chest x-ray clearly illustrated a right-sided aortic arch, pulmonary artery bay, elevated cardiac apex typical of right ventricular enlargement, but an otherwise normal cardiac silhouette (Figure 1). ECHO showed a large (8mm) perimembranous VSD with a pure right-to-left shunt, an overriding aorta with a right-sided arch, and several collateral branches from the descending aorta to the pulmonary arteries (MAPCAs). The branch pulmonary arteries (PA) were difficult to visualise. A closed Ductus Arteriosus was identified, and there was no coarctation of the aorta. There was a small patent foramen ovale with a predominantly left-to-right shunt. The right ventricle was dilated, with trivial tricuspid regurgitation. Both ventricles displayed good systolic function. A CT angiogram (Siemens, Flash Dual Source, Germany) with iopromide (Ultravist©) contrast was utilised to determine the pulmonary arteries and the MAPCAs more accurately (Figure 2). The patient then successfully underwent surgery to establish antegrade connection between her right ventricle and pulmonary arteries.

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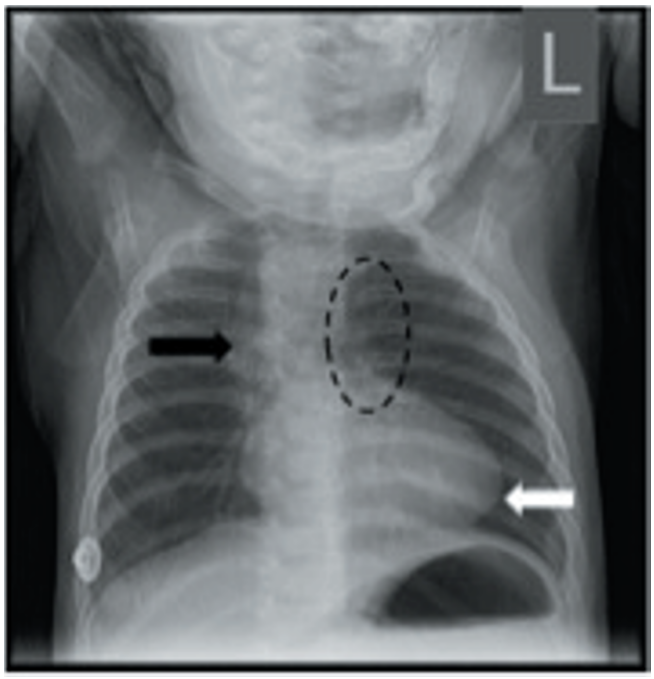


**Figure 1**
An AP erect chest xray of the patient showing an upward tilted cardiac apex, suggesting RV enlargement (white arrow); a right-sided aortic arch, indenting the trachea on the right side (black arrow); pulmonary bay suggesting small or absent pulmonary arteries (dotted oval).

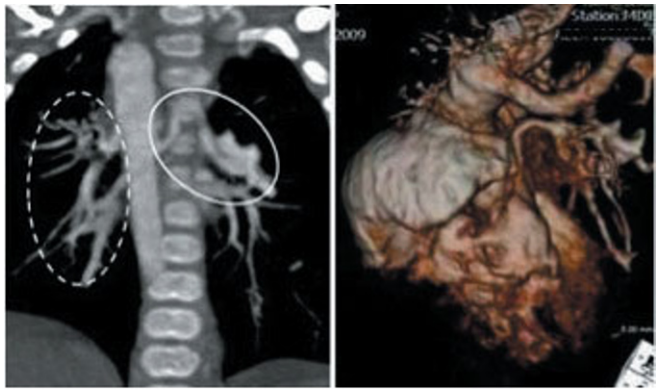

**Figure 2**
A CT angiogram of the patient demonstrating a right-sided aorta and MAPCAs (left) and a 3D reconstruction of the CT examination (right). A MAPCA arises from the left side of the aorta, loops superiorly then inferiorly and supplies the left upper lobe (solid white oval); Two MAPCAs arise from the right side of the aorta and supply parts of the right middle and lower lobes (dotted oval).

With the continuous advancement in imaging modalities, clinicians have at their disposal a number of reliable imaging modes for diagnosis. This is especially true in the setting of congenital heart disease. Since the advent of cardiac catheterisation and echocardiography in the 1960s and 1980s respectively, several other imaging tools have emerged, including Magnetic Resonance Imaging (MRI) and CT angiography. And while each have their own advantages, they also come with their respective shortcomings, which should be taken into account, and minimised. This may include using a combination of imaging methods to complement one another.

With its convenience of use, well-established echocardiographic-morphological correlations, and non-invasive nature, ECHO is always the first-line diagnostic investigation in patients with suspected CHD (2). The additional Doppler techniques (Figure 3), which allowed visualisation and flow velocity quantification, have enhanced the diagnostic accuracy of the ECHO (3). As outlined in our case report above, much of the anatomical and physiological detail could be determined using ECHO. Currently a significant number of CHD cases are referred directly for interventions based chiefly on an echocardiographic diagnosis only (4). While ECHO remains as the most widely used imaging technique in the diagnosis of CHD, it does have its limitations, particularly in complex congenital anomalies involving extracardiac structures, such as small central pulmonary arteries, peripheral pulmonary arteries, the cervical vessels and pulmonary veins (2, 4). This case demonstrates the difficulties in determining the detailed anatomy of hypoplastic central pulmonary arteries and the size, number and distribution of MAPCAs by echocardiography.

Given the limitations of echocardiographic diagnosis in certain CHD cases, other imaging modality may be needed to provide additional information. Cardiac MRI is the gold standard technique for depicting aortic anomalies and relevant collateral vessels (5). Furthermore, the ability of MRI to assess volume and mass of peculiar ventricular shapes, which may be a feature of CHD, with optimal accuracy, is especially desirable (6). Compared to other imaging modalities such as cardiac catheterisation and CT, MRI studies do not involve exposure to ionising

radiation. As such, accepted pediatric indications for cardiac MRI include: segmental depiction of cardiac morphology, detailed evaluation of aortic and other major vessel and systemic venous anomalies, quantification of shunts, stenoses, and regurgitation, and post-operative study (6-9). However, certain drawbacks do limit the utilisation of MRI. Study time is a key limitation for MRI, with a typical congenital cardiac study using a 1.5 T magnet requiring 45-60 min (10). And while this is less of an issue with adult patients, children may require sedation, or general anaesthesiae, which may confer additional risks (11). Furthermore, there are technical barriers to sequence optimisation in the use of MRI particularly in the young paediatric age group; which include: fast heart rate requiring high temporal resolution for optimal ventricular volume and flow assessments, and small cardiac size and vessels which require great spatial resolution (12). Another potential challenge that one should keep in mind is the resuscitation of a critically ill patient in the confines of the magnet, which require removal of the patient from the scanning room (13). Although diagnostic cardiac catheterisation with selective angiography in each MAPCA provides precise diagnostic information, it also carries a significant risk (14). Thus, given the young age and clinical state of our patient, we felt that although MRI and cardiac catheterisation would have provided an accurate anatomical depiction, the benefits did not justify the potential risks, and CT was a more suitable imaging option.

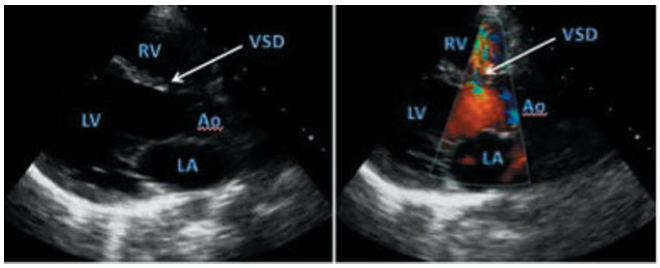


**Figure 3**
An echocardiographic image demonstrating perimembranous VSD (left), enhanced with Doppler function (right). LV=left ventricle; LA=left atrium; RV=right ventricle; Ao=aortic outlet; VSD=ventricular septal defect

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The development of CT technology, with the multidetector scanners and 16-, 32-, 64- and 128-detector technology has allowed extremely fast image acquisition (10 ± 2s) without significant compromise to image quality (4, 10). It provides high spatial resolution and detailed 3D reconstruction

of extra-cardiac anatomical structures, such as pulmonary vessels e.g. pulmonary sling (Figure 4), aortic arch anomalies e.g. right-sided aortic arch (Figure 5), and coronary arteries (15-16). In addition to its minimal invasiveness, MDCT has also been shown to be more superior than conventional cardiac catheterization in delineating MAPCAs, in particular for identifying segmental pulmonary blood flow (17). The difficulties in precise diagnosis of right-sided heart anomalies potentially can be overcome by CT (2). The short acquisition times of CT are advantageous over MRI since it effectively provides a means for accurate, detailed imaging without the need for sedation or general anaesthesia in paediatric patients (10), even for small infants. Nevertheless, the benefits of CT need to be weighed up against the associated exposure to ionizing radiation and contrast agents. The risk of cancer associated with radiation exposure needs to be considered, especially in children, who are at greater risk given they are inherently more radiosensitive (18). This may be minimised by adjusting the dose for the child's weight to target the minimum possible radiation exposure, while maintaining diagnostic image quality (19).

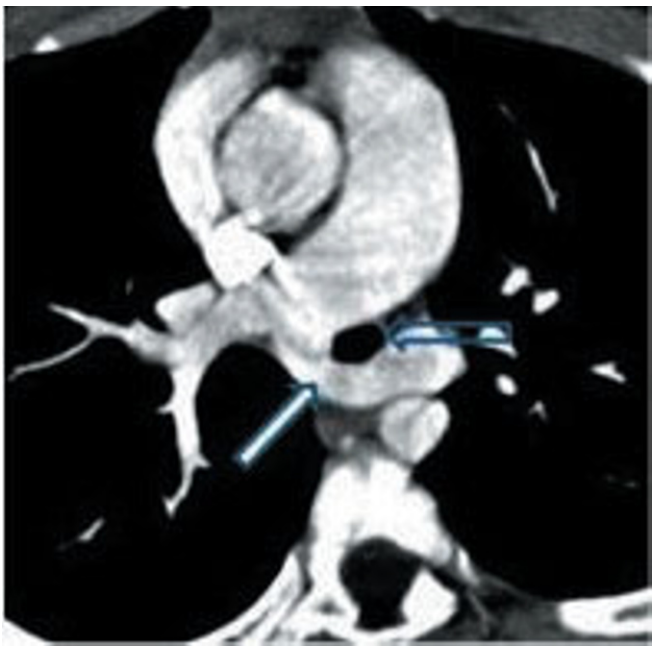

**Figure 4**
A CT angiogram of a classic pulmonary sling, showing an anomalous left pulmonary artery (solid arrow) coursing to the left hemithorax posterior to the trachea (clear arrow).

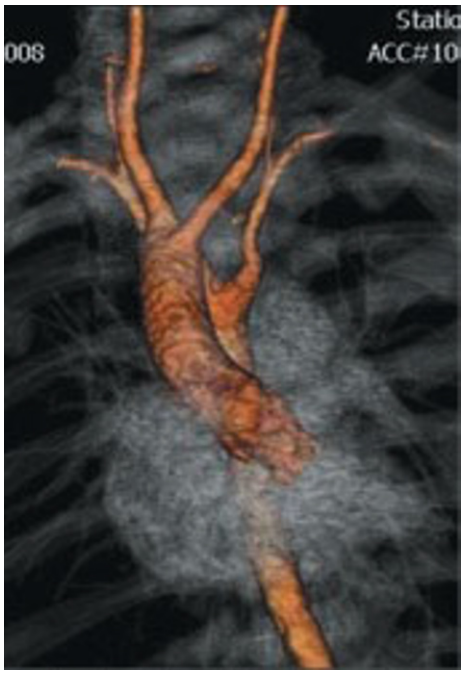


**Figure 5**
A CT reconstruction of a right-sided aortic arch.

## Conclusion

With the increasing number of imaging modalities available in the practice of paediatric cardiology, clinicians need to be familiar with the benefits and limitations of each technique, and always consider the options to complement, or substitute, one tool with another, to provide optimal patient management.



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