Application of Vessel Navigator™ fusion imaging software in a complex transcatheter palliation of Tetralogy of Fallot with pulmonary atresia

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ABSTRACT

Extreme pulmonary artery hypoplasia in cyanotic malformations precludes palliative surgeries. When aortopulmonary collaterals (APC) in such patients are also hypoplastic, their unifocalization to create a neopulmonary vasculature is also hampered. Stent angioplasty of the outflow or collateral arteries may reduce hypoxia but is challenging in tortuous and atretic tracts. Fusion imaging overlays anatomical data from computed tomography during adult structural interventions, but its use is not often reported in young children with complex cyanotic malformations. This report shows utility of fusion imaging in pulmonary atresia with extremely hypoplastic pulmonary arteries and stenotic APC to guide stenting of outflow tract and collaterals.

Keywords: Aortopulmonary collaterals, computed tomography, pulmonary valve perforation, right ventricular outflow tract, stent angioplasty

INTRODUCTION

Surgical treatment of Tetralogy of Fallot with pulmonary atresia (TOF-PA) is standardized in patients with adequate pulmonary arteries.[1] When severely hypoplastic pulmonary arteries preclude even an aortopulmonary shunt, surgical techniques rely on unifocalizing aortopulmonary collaterals (APC) to create a neopulmonary vasculature. Options are restricted when both pulmonary arteries and mediastinal APC are minuscule in size.[2] Tortuosity and acute angulations of APC challenge their transcatheter stent angioplasty. Vessel Navigator™ software (Koninklijke Philips N.V., Best, Netherlands) overlays three-dimensional vascular anatomical information from computed tomography (CT) on fluoroscopy that guides to maneuver tortuous stenotic vessels for vascular interventions.[3,4] We report a first use of this novel technique in pediatric cardiology to guide the perforation of a thick atretic pulmonary valve, stent dilatation of the right ventricular outflow tract (RVOT), and stent angioplasty of two stenotic APC.

CASE REPORT

A 4-year-old boy weighing 12 kg diagnosed to have TOF-PA presented with breathlessness relieved by squatting along with motor developmental delay owing to severe hypoxia. Speech and intelligence were normal, and there were no syndromic features. Oxygen saturation was 65% at rest. There were no murmurs. Chest X-ray showed severe pulmonary oligemia without cardiomegaly. Hemoglobin was 19.1 g/dl, and hematocrit was 60%.

How to cite this article: Sivakumar K, Mumtaz ZA, Sagar P. Application of Vessel Navigator™ fusion imaging software in a complex transcatheter palliation of Tetralogy of Fallot with pulmonary atresia. Ann Pediatr Card 2022;15:187-91.

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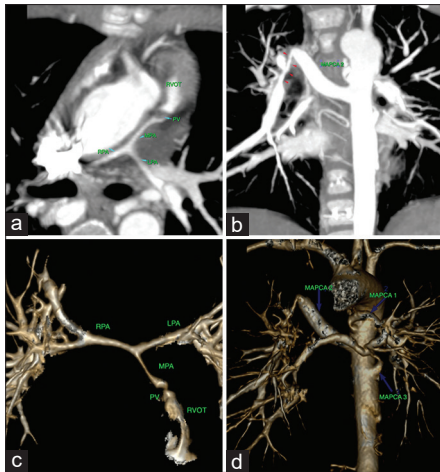E-mail: drkumarsiva@hotmail.com
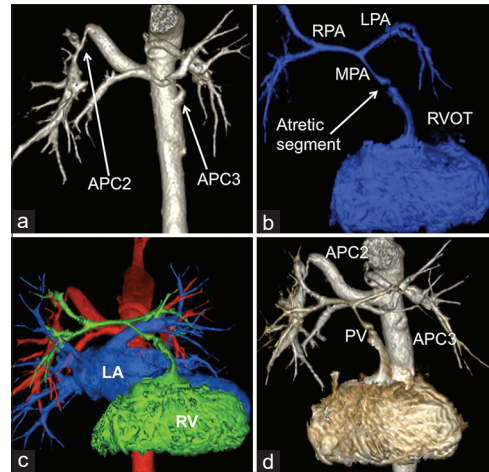
Figure 1: Axial (a) and coronal (b) computed tomography images show a blind right ventricular outflow tract ending in an atretic pulmonary valve. The main, right, and left pulmonary arteries are markedly hypoplastic measuring 1 mm each. The aortopulmonary collateral has long segments of stenosis (red arrows). Volume rendering of the outflow tract (c) and aortic collaterals (d) shows three collaterals numbered 1–3, the last two are stenotic. RVOT: Right ventricular outflow tract, PV: Pulmonary valve, MPA: Main pulmonary artery, RPA: Right pulmonary artery, LPA: Left pulmonary artery, MAPCA: Major aortopulmonary collateral artery

Figure 2: Reconstructed images (a) shows long segmental stenosis of second (APC2) and third (APC3) collaterals. An atretic pulmonary valve is seen between outflow (RVOT) and pulmonary trunk. (b) Color rendering (c) combines both these structures along with left atrium. In order to facilitate fusion with fluoroscopy, the left atrium is removed (d). APC: Aortopulmonary collateral, RVOT: Right ventricular outflow tract, PV: Pulmonary valve, MPA: Main pulmonary artery, RPA: Right pulmonary artery, LPA: Left pulmonary artery

Echocardiogram failed to identify any pulmonary arteries. CT demonstrated three small APCs with severe stenosis of mediastinal part of the second and third APC that measured 3–4 mm beyond the stenosis [Figure 1]. There was a 2-mm thick atretic pulmonary valve at the end of the pulmonary trunk; mediastinal right and left pulmonary arteries measured 1 mm each. Multidisciplinary team decided against surgical shunts and unifocalization, as vessels were hypoplastic. After informed consent, a plan was made to recanalize the atretic pulmonary valve, stent the RVOT, and stent the APC [Figure 2].

Vessel Navigator™ software segmented the APC as well as the atretic RVOT and pulmonary arteries [Figure 3]. While a very steep cranial projection that is unachievable in fluoroscopy profiled the atretic RVOT, an anteroposterior projection showed the first two APC, and a left anterior oblique steep caudal projection profiled the third APC. Mechanical ventilation ensured airway protection, adequate oxygenation, and lack of patient mobility aided fusion. The first communicating APC at D4 level from the anterior surface of aorta supplied dorsal segment of the left lower lobe. The second APC at D5 level from right surface that faintly communicated to the central pulmonary arteries supplied the right mid and lower zones with stenosis after a tortuous bend. The third noncommunicating APC at D6 level from the anterior surface supplied the left upper lobe and right lower lobe with a tightly stenosed long segment immediately after its origin.

The blind atretic RVOT was engaged using the image overlay in two orthogonal projections with confirmatory check injections [Figure 4]. The atretic membrane was perforated using a Conquest Pro20 guidewire (Asahi Intecc, Aichi, Japan) along the line of pulmonary trunk, and the wire position in the branch pulmonary artery was confirmed by a communicating APC injection. The atretic segment was balloon dilated with a 1.25-mm coronary balloon before stenting with 4.5 mm × 30 mm Resolute Onyx (Medtronic, Minneapolis, MN, USA). The first APC supplying only the dorsal segment of left lower lobe was not significantly narrowed. The long segment stenosis in the second APC was stented with another 4-mm Resolute Onyx. The third APC supplying both lungs had a long narrowed segment overlying the descending aorta. A left anterior oblique caudal projection guided by fusion imaging aided its stenting with another 4-mm Resolute Onyx [Figure 5].

The fluoroscopic time was 57 min, and the dose area product was 32.7 Gy.cm². The contrast volume was 40 ml. The oxygen saturation improved to 90% with a marked relief of symptoms, improved motor activity, and an easily audible outflow tract systolic murmur. After 3-month follow-up on aspirin and clopidogrel, the oxygen saturation was maintained, effort tolerance had considerably improved and CT confirmed stent patency [Figure 6].

DISCUSSION

A key determinant for success in the management of cyanotic malformations is the adequacy of the pulmonary
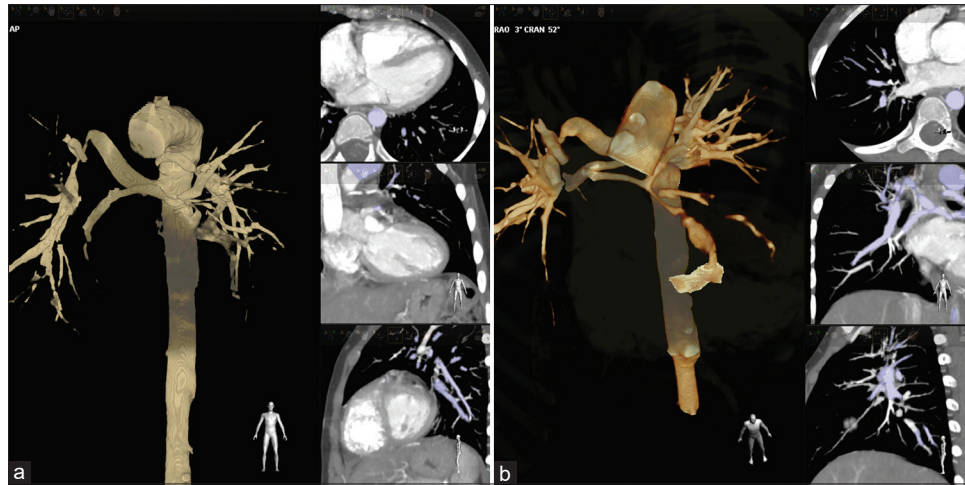
Figure 3: Segmentation of aortopulmonary collaterals (a) on multiple orthogonal axial, coronal and sagittal planes aids in reconstruction of the aorta with the three collaterals as well as the addition of right ventricular outflow and branch pulmonary arteries (b)
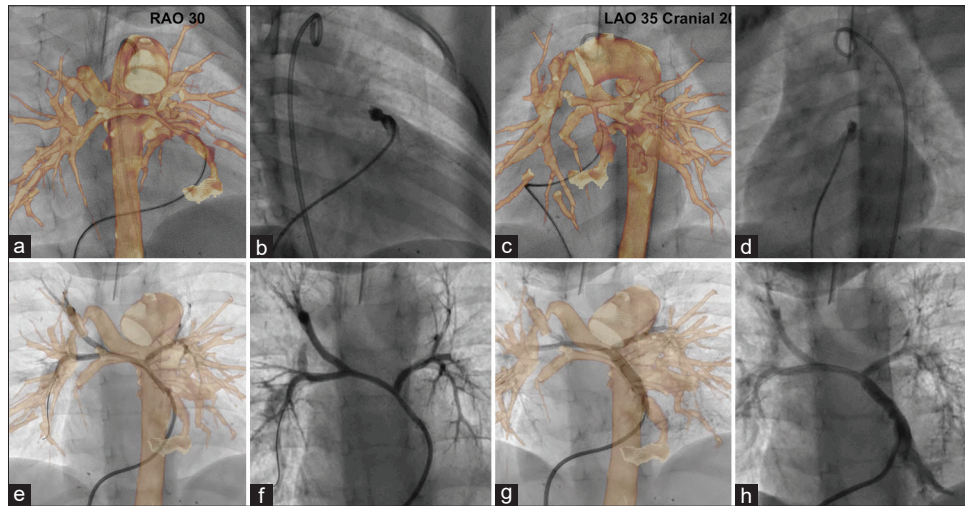

Figure 4: Fluoroscopic images with (a and c) and without (b and d) fusion in orthogonal right anterior oblique (a and b) and left anterior oblique projections (c and d) guide engagement of the atretic pulmonary valve with a catheter. After perforation of the atretic valve and balloon dilatation, angiogram shows small hypoplastic pulmonary arteries smaller than the catheter (e and f). After outflow tract stenting, images (g and h) demonstrate the stent position fused with the overlay

arteries.[1] An extremely hypoplastic vessel measuring 1–2 mm is not suited for even a palliative shunt. Balloon and stent angioplasty of a patent RVOT may improve the flow into hypoplastic pulmonary arteries.[5] Recanalizing atretic pulmonary valve is commonly performed in PA with intact ventricular septum, but rare in TOF-PA as the atretic segment is often long.[6]

CT delineated the RVOT anatomy precisely in this patient with a 2-mm thick atretic membranous plate. Fusion imaging aided to advance the catheter and abut it against the atretic segment. The heavy tip load of 20 g of Conquest Pro wire was advanced in an accurate direction toward the pulmonary trunk. RVOT perforation into the pericardium was potentially avoided by the fusion, which also guided the subsequent stent positioning.[6]

Stenotic APC is an additional interventional target to improve hypoxia and may contribute to the growth of communicating central pulmonary arteries. Their eccentric aortic origin and tortuous course may challenge the intervention. Resistance to balloon dilatation and restenosis make them unattractive targets.[7]

While the first APC had a very restricted distribution to the dorsal segment of the left lower lobe, the second APC had a long stenotic segment before arborizing to the entire right lung. Fusion imaging provided the guidance for precise cannulation of the collateral, advancing the guidewire and positioning the stent. Fusion was best utilized in the third tortuous APC overlying the descending aorta, profiled best on a left anterior oblique caudal projection that guided the procedure.[3] Conventional angiography of an unknown vessel such as APC often involved trial
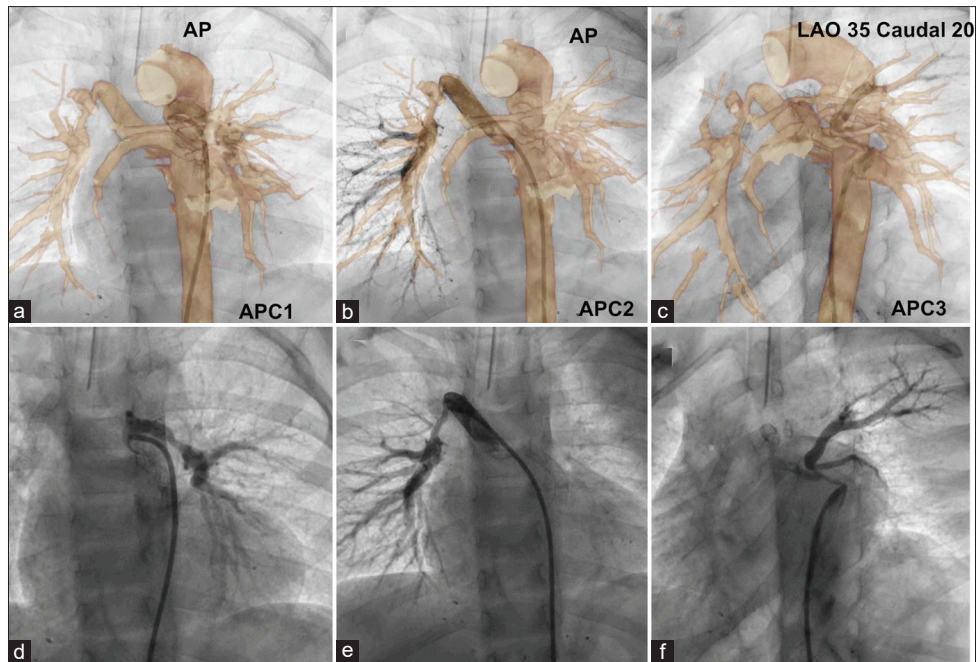
Figure 5: Angiography is performed in anteroposterior projection for delineating first (a) and second (b) collaterals, but in left anterior oblique caudal projection for the third collateral (c). Images without fusion demonstrate limited arborization to left lower lobe from the first collateral (d), many segments of right lung from a stenotic second collateral (e) and to both lungs from another stenotic third collateral (f) artery. APC: Aortopulmonary collateral
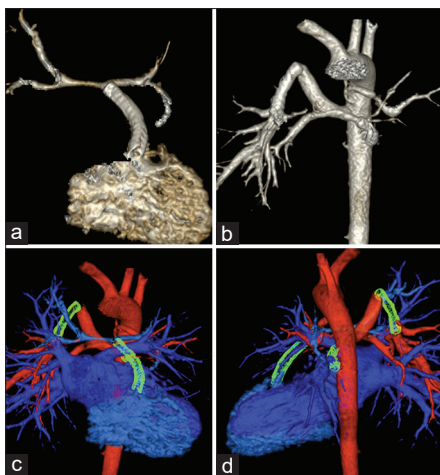
Figure 6: Volume rendered computed tomographic images show stented outflow (a) and stented aortopulmonary collaterals (b). Color rendering viewed from front (c) and behind (d) demonstrating the three dimensional perspective of the three stents

and error multiple orthogonal views with excessive use of radiation and contrast media.[8]

Extreme hypoxia and marked hypoplastic pulmonary arteries did not permit a staged approach of initial perforation and RVOT stenting, followed by a deferred decision on APC stenting. Even though RVOT stenting provided a forward flow promoting arterial growth, extreme hypoplastic arteries measuring 1 mm might not increase their size significantly in the short term. In addition, an arterial access that was taken initially for the selective APC angiogram was used for their stenting

in the same sitting, thereby avoiding repeated arterial punctures on immediate follow-up.

Improved precision in adult structural interventions such as left atrial appendage occlusion, percutaneous valve implantation, transapical puncture, coarctation stenting, and others was shown earlier using fusion imaging.[9] However, this technology was sparingly used in young children, especially in interventions involving perforation of an atretic pulmonary valve in the setting of TOF-PA and stenting of narrow tortuous APC.[3,4] Anatomy was crucial for a guidewire perforation through the narrow infundibulum with very little margin of error.[6] Fusion showed the trajectory to advance the wire from the right ventricle into the pulmonary trunk. It also permitted precise positioning of the stent without orthogonal angiography. Even though selection of fluoroscopic projection guided by fusion reduced radiation duration and contrast volumes, our complex long procedure involved three major interventions in one single sitting.[4,10]

CONCLUSIONS

Complex vascular interventions including perforation of atretic pulmonary valve, stent angioplasty of severely hypoplastic outflow tracts, and tortuous stenotic APC may be guided by fusion imaging utilizing CT even in young children. It may aid precision, reduce multiple angiograms, selection of the most appropriate projection, and ultimately reduce radiation dose. Future attempts to

use fusion imaging in young infants who require complex interventions may establish its utility in such patients who need it the most.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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