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Brief Report

Hemiazygos vein "steal hypoxic-syndrome" after hemi-Fontan operation: comprehensive four-dimensional flow magnetic resonance visualisation

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Abstract A 2-year-old boy with hypoplastic left heart syndrome and previous hemi-Fontan palliation surgery was referred for cardiovascular magnetic resonance evaluation because of progressive cyanosis. This case report illustrates the advantages of non-invasive four-dimensional magnetic resonance imaging for comprehensive identification and quantification of venovenous collaterals in patients with palliated hemi-Fontan staged surgery.

Keywords: Phase-contrast flow; hypoplastic left heart syndrome; venovenous collaterals

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SINGLE-VENTRICLE PALLIATION WITH A SUPERIOR cavopulmonary anastomosis may result in elevated pressure in the superior caval vein, which may result in a raised transpulmonary pressure gradient. In one-third of the patients, this may lead to the development of venovenous collaterals mainly in the azygos and hemiazygos system, but also in other any small tributary vein normally draining to the superior or inferior caval vein.[1] Such venovenous collateral channels can lead to profound systemic desaturation and have been described previously as the "steal hypoxic-syndrome".[2]

Careful evaluation for venovenous collaterals is mandatory and usually relies on non-invasive imaging modalities: echocardiography and magnetic resonance imaging. Unfortunately, owing to its anatomical variability and small size, identification of venovenous collaterals and, more important, quantification of the "stolen" collateral flow is challenging. As this information informs further management decisions, invasive diagnostic cardiac catheterisation has hitherto often been required.[3] We present a non-invasive four-dimensional phase-contrast flow magnetic resonance technique to precisely evaluate the anatomy and quantify the venovenous collateral flow in a patient with palliated hypoplastic left heart syndrome and contrast the potential benefits of four-dimensional over other imaging techniques.

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Case report

We present the case of a 2-year-old boy (8.5 kilograms of body weight) with an antenatal diagnosis of hypoplastic left heart syndrome, aortic atresia. The patient underwent a Norwood Stage-I surgical palliation soon after birth and a hemi-Fontan superior cavopulmonary anastomosis at 6 months of age – oxygen saturations at hospital discharge 84–94%. Only 6 months after the hemi-Fontan operation, he became increasingly cyanotic with oxygen saturations between 70% and 75% by pulse oxymetry. Echocardiography suggested the existence

of a venovenous collateral adjacent to the superior caval vein, but the distal course of this venovenous collaterals could not be evaluated echocardiographically. The patient was therefore referred for magnetic resonance imaging study. The scan was performed under general anaesthesia using a 1.5 T Intera Magnetic Resonance scanner (Philips Medical Systems, Best, the Netherlands). Routine protocols included anatomic evaluation by three-dimensional contrast-enhanced magnetic resonance angiography (repetition time/echo time = 4.8/1.5 milliseconds, spatial resolution = 1.3 × 1.3 × 1.5 millimetres) and haemodynamic flow assessment with conventional two-dimensional phase-contrast flow (repetition time/echo time = 4.4/2.4 milliseconds, spatial resolution = 2.5 × 2.5 × 8 millimetres, temporal resolution = 17 milliseconds, maximal velocity encoded = 60–250 centimetres per second, acquisition time 2:10 minutes) in five planes to measure the through plane flow in the right and left pulmonary arteries, superior and inferior caval vein, and aorta. Two-dimensional flow acquisition to quantify the through plane flow in the venovenous collaterals was not possible because of the relatively small vessel size and tortuous course. However, a single four-dimensional sequence including the whole mediastinum was acquired (repetition time/echo time = 4.8/2.7 milliseconds, spatial resolution = 2.5 × 2.5 × 2.5 millimetres, temporal resolution = 25 milliseconds, maximal velocity encoded = 250 centimetres per second, acquisition time 11:40 minutes).

Detailed post-processing reconstruction of the contrast-enhanced magnetic resonance angiography confirmed a dilated hemiazygos system and delineated the distal venous course and, importantly, did not identify any major aortopulmonary collateral (Fig 1a). Despite these clear anatomical data, this sequence does not provide haemodynamic flow information. It was therefore not possible to clarify whether the hemiazygos vein had a normal venous flow draining towards the superior caval vein, or had reversed flow from the superior to the inferior caval vein, thus functioning as a venovenous collateral channel. In summary, contrast-enhanced angiography sequence could not quantify the amount of any "stolen" venovenous collaterals flow.

However, retrospective evaluation of the four-dimensional data allowed a comprehensive quantification of all intrathoracic vessels (Fig 1b). Flow in the hemiazygos vein was significant (0.6 litre per minute per square metre, 17% of the aortic systemic flow) with reversal of direction towards the inferior caval vein (see Fig 2 and online Supplementary Video S1). In this patient, the hemiazygos vein functioned as a venovenous collateral, thereby reducing the effective pulmonary blood flow by 37%.

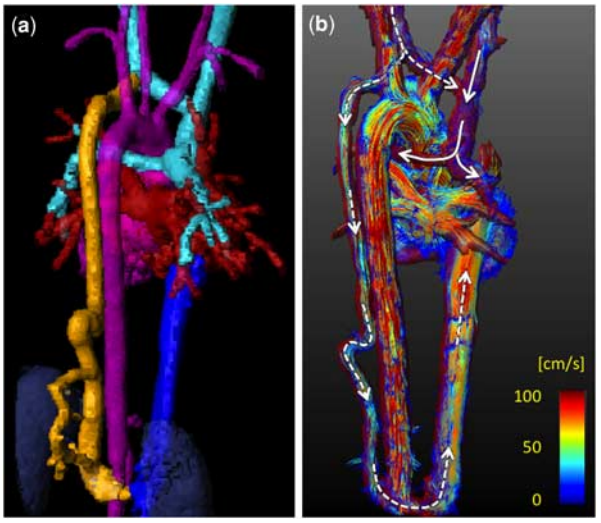

Figure 1.
*Comparison of contrast-enhanced angiography (**a**) and four-dimensional (**b**) magnetic resonance imaging. (**a**) Contrast-enhanced magnetic resonance angiography, posterior view: The hemi-Fontan anatomy and the presence of the hemiazygos system are precisely shown, but no haemodynamic information about the flow stream is provided. Superior caval vein and pulmonary arteries (shaded blue), hemiazygos vein (gold), inferior caval vein (navy blue), pulmonary veins (red), single ventricle and aorta (purple). (**b**) The four-dimensional flow provides anatomical and also haemodynamic information in the hemi-Fontan circulation (solid arrows) and the hemiazygos steal syndrome, with reversal downstream blood flow draining into the inferior caval vein (dotted arrows). See also supplemental movie.*

Discussion

The azygos and hemiazygos vein system is formed by a paired longitudinal network of small veins connecting the inferior caval vein, abdominal and thoracic wall veins to the superior caval vein. Following superior cavopulmonary anastomosis surgery – in this case a hemi-Fontan – an increase in the superior caval vein pressure may lead to a reversal of flow in the azygos and hemiazygos vein system, reduction in pulmonary blood flow, and clinical cyanosis. Some centres advocate routine ligation and division of the azygos vein at the time of the superior cavopulmonary anastomosis surgery. This does not, however, prevent the development of venovenous collaterals in the hemiazygos vein or other vascular territories. Worsening cyanosis always mandates further investigations to exclude venovenous collaterals, but because of the variability and retro-mediastinal location, identification and quantification is challenging.

Although diagnostic cardiac catheterisation is considered to be the gold standard for investigation of venovenous collaterals, alternative methods are preferred because of the invasive nature of this procedure and the risk of radiation exposure.[4]

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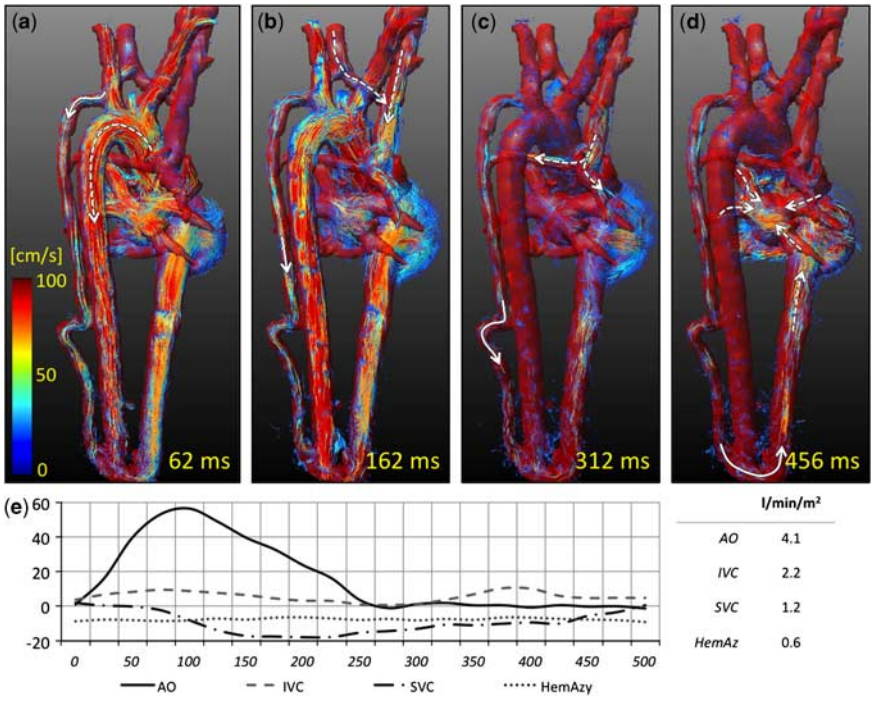

Figure 2.
*Four-dimensional flow quantification. (**a–d**) Four-dimensional velocity-coded particles traces at different time points of the cardiac cycle: 62, 162, 312, and 456 milliseconds. Velocity expressed as centimetres per second. (**e**) Four-dimensional direct quantification of individual vessels: ascending aorta (AO), inferior caval vein (IVC), superior caval vein (SVC), and hemiazygos vein (HemAz). The plot shows the velocity profile (centimetre per second) of the different vessels over time (millisecond) in one averaged cardiac cycle. The flow in the ascending aorta and inferior caval vein have a feet to head direction and are coded as positive velocity. The flow in the superior caval vein and hemiazygos vein have a downstream head to feet direction and are coded as negative velocity. The net-indexed flow in each vessel expressed as litre per minute per square metre is shown on the right panel.*

Echocardiography may identify collateral vessels but does not provide sufficient anatomical detail to delineate the full vessel anatomy. Computed tomography can provide this anatomical information, is also non-invasive, but poses a radiation risk.[5] Contrast-enhanced magnetic resonance provides detailed cardiovascular anatomy with no radiation but does not measure flow. Addition of two-dimensional phase-contrast flow to contrast-enhanced angiography can quantify the blood flow, but in cases where the anatomy is complex it is time consuming, requiring several sequences that may be difficult to plan because of vessel morphology and orientation.

Four-dimensional magnetic resonance imaging is a recently developed technique, which has shown great potential for application in patients with congenital heart disease.[6,7] Four-dimensional magnetic resonance can be planned as a time-resolved three-dimensional volume data set to include the whole intrathoracic cardiovascular system. Even in the presence of unexpected findings, careful retrospective analysis allows quantification of any additional thoracic vessel without adding extra scanning time. In this case, four-dimensional required similar time to the five two-dimensional flow acquisitions (11:40 versus 10:50 minutes), but importantly also included additional vessels information. To our knowledge, this is the first report to quantify non-invasively the flow in a venovenous collateral causing the hemiazygos "steal hypoxic-syndrome". The resulting additional morphological and physiological data obtained non-invasively without radiation will aid decision making before further catheter or surgical intervention in patients with venovenous collaterals. The four-dimensional technique may be considered as an alternative to conventional two-dimensional flow sequence, particularly in complex cases where multiple vessel flow estimations are required.

Supplementary materials

For supplementary materials referred to in this article, please visit http://dx.doi.org/doi:10.1017/S1047951112000248

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