Imaging findings in Steidele's complex

Arvind Kandoria, Rajeev Bhardwaj, Neeraj Ganju, Kunal Mahajan

Indira Gandhi Medical College, Shimla, Himachal Pradesh, India

Correspondence to
Dr Kunal Mahajan,
kunalmahajan442@gmail.com

Accepted 17 May 2017

DESCRIPTION

Steidele's complex is an eponym to interrupted aortic arch (IAA), which is characterised by complete anatomic discontinuity between the aortic arch and the descending aorta (DA). It is a very rare congenital anomaly that occurs in 19 per million live births.[1] Based on the site of interruption, it is classified into three types: type A (30%–40%) when it occurs beyond the left subclavian artery, type B (51%–70%) when it occurs between the left carotid artery and the left subclavian artery and type C (1%–5%) when it occurs between the brachiocephalic artery and the left common carotid artery.[2] DA originates from the pulmonary artery through a patent ductus arteriosus (PDA). IAA is invariably associated with a ventricular septal defect (VSD) in majority of cases. IAA is associated with a high mortality rate of 75% at 1 month and 90% at 1 year, in the absence of operative intervention.[3] We describe a 3-year-old girl who presented with failure to thrive. There was no history of recurrent respiratory tract infections, heart failure and cyanosis. There was no disparity in peripheral pulses or differential cyanosis found on clinical examination (figure 1); however, saturation in upper limbs was 94% in comparison to 80% in lower limbs, recorded by pulse oximetry. Cardiac examination revealed a prominent right ventricular impulse and a grade 2 ejection systolic murmur in the pulmonary

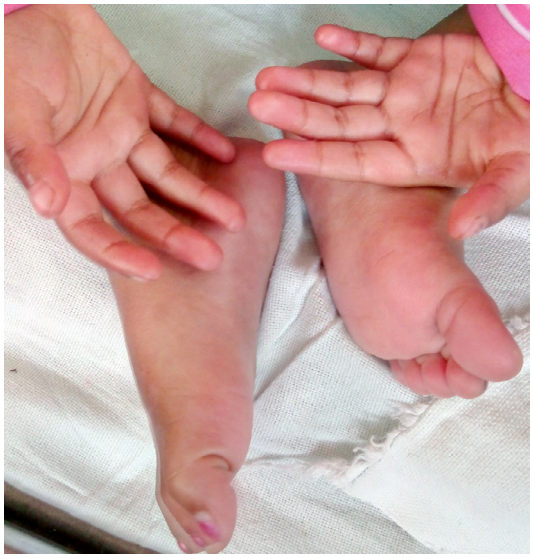


Figure 1 Clinical image revealing the absence of differential cyanosis.

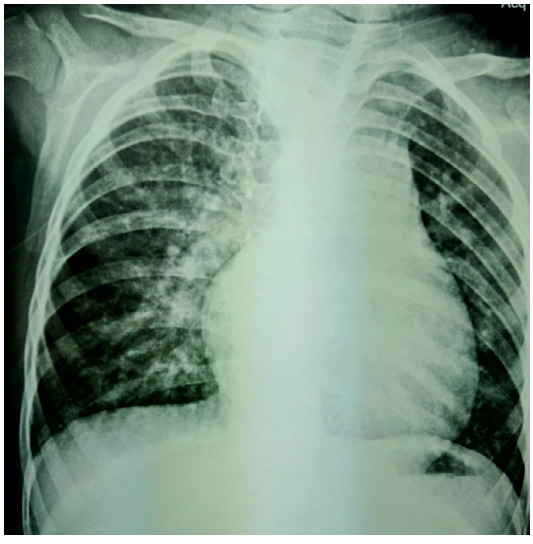


Figure 2 Chest X-ray showing increased pulmonary blood flow in the right lung but not in the left lung. MPA and RPA are prominent. Note that the apex is the right ventricle type and the right atrium is enlarged. MPA, main pulmonary artery; RPA, right pulmonary artery.

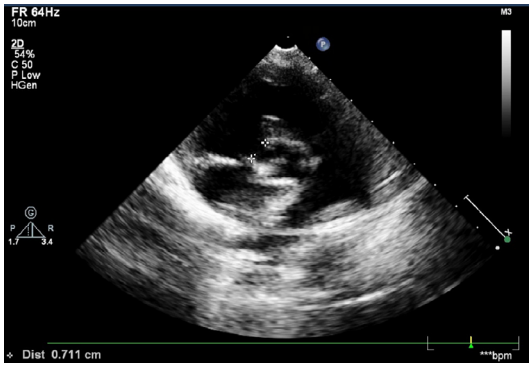


Figure 3 Transthoracic echocardiogram-parasternal short axis view showing a 7 mm perimembranous ventricular septal defect.

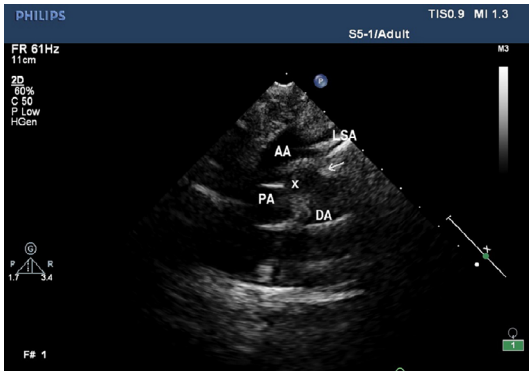


Figure 4 Transthoracic echocardiogram-suprasternal view showing an interrupted aortic arch (arrowhead) distal to the LSA. Note that the PA, ductus (x) and DA are in continuity. Also seen is the dilated pulmonary trunk in comparison to the ascending aorta. DA, descending aorta; LSA, left subclavian artery; PA, pulmonary artery.

To cite: Kandoria A, Bhardwaj R, Ganju N, *et al*. *BMJ Case Rep* Published Online First: [*please include* Day Month Year]. doi:10.1136/bcr-2017-219592

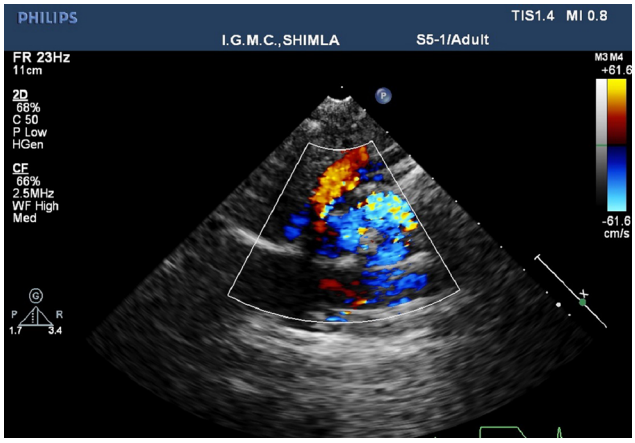
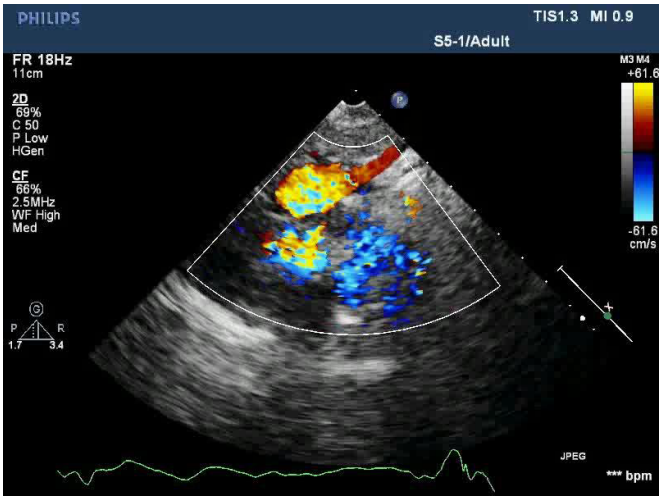


Figure 5 Suprasternal view, colour Doppler imaging, reveals colour flow continuity between PA and descending artery via the patent ductus. Colour flow demonstration is also seen from the arch into the LSA but not into the DA. DA, descending aorta; LSA, left subclavian artery; PA, pulmonary artery.



video 1 Transthoracic echocardiography-suprasternal view demonstrating colour flow across the PA into the DA through the patent ductus. Note that there is colour flow from the arch into the LSA but not into the DA. DA, descending aorta; LSA, left subclavian artery; PA, pulmonary artery.

Learning points

- Steidele's complex is a rare anomaly with high mortality rate of around 75% at 1 month in untreated cases, which coincides with the ductal closure. Mortality is 90% at 1 year without surgical intervention. Late presentation is less commonly reported in children, adults and even the elderly.
- On clinical examination, disparity in peripheral pulses or saturation and the absence of any significant murmur in the presence of a prominent right ventricular impulse should raise the suspicion of Steidele's complex.
- Diagnostic clues on echocardiography include smaller calibre ascending aorta in comparison to pulmonary trunk and a continuity between pulmonary artery, patent ductus arteriosus and the descending aorta.

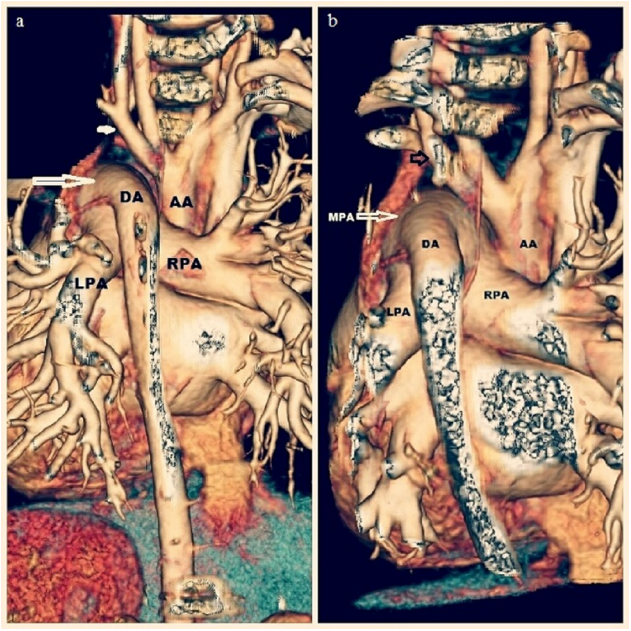


Figure 6 MDCTA images (**A and B**) showing interruption of aortic arch after the origin of LSA (smaller arrows). Note the MPA (larger arrows) giving origin to DA followed by origin of LPA and RPA. Interestingly, LPA is smaller in calibre to RPA probably because of differential streaming of blood into DA and RPA. DA, descending aorta; LPA, left pulmonary artery; LSA, left subclavian artery; MDCTA, multidetector CT angiography; MPA, main pulmonary artery; RPA, right pulmonary artery.

area. ECG revealed right atrial enlargement and right ventricular hypertrophy. The chest X-ray was suggestive of dilatation of the main pulmonary artery (PA) and the right descending PA. The left PA was not dilated and increased pulmonary blood flow was marked in the right lung, but not in the left lung (figure 2). Echocardiogram showed a perimembranous VSD with a left to right shunt and a dilated main PA (figure 3). IAA was suspected on the suprasternal window imaging (figure 4). The main PA was in continuity with the DA through PDA (figure 5, video 1). These findings were confirmed on CT imaging. Interestingly, CT also demonstrated preferential dilatation of the main and right PA, while the left PA was normal in size (figure 6). This must be due to preferential streaming of blood into the DA and right PA. The patient underwent successful surgical repair.

Contributors AK, RB and KM made the diagnosis and performed the complete investigations and were involved in the management of the patient. KM wrote the manuscript. NG performed the literature search. AK and RB corrected the manuscript and gave conceptual advice. All authors read and approved the final version of the manuscript.

Competing interests None declared.

Patient consent Consent obtained from guardian.

Provenance and peer review Not commissioned; externally peer reviewed.

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