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Letter to the Editor

Sir,

Re: **Right isomerism, pulmonary atresia, and major aorto-pulmonary collateral arteries.**

We read with interest the recent report by Kumar et al.[1] on the association of major aorto-pulmonary collateral arteries with right isomerism. Major aorto-pulmonary collateral arteries, with or without discontinuity of the intrapericardial pulmonary arteries, are quite rare in patients with right isomerism. They are seen far more frequently in the subset of children with pulmonary atresia with ventricular septal defect in which the intracardiac anatomy is that of tetralogy of Fallot.[2] Aorto-pulmonary collateral arteries, in fact, are reported to exist in up to one-tenth of cases of right isomerism, or visceral heterotaxy with asplenia.[3,4] Even in children with pulmonary atresia and right isomerism, the collateral arteries are not as rare as in patients with pulmonary atresia when the ventricular septum is intact.[5,6] Arterial supply to the lungs through bilateral arterial ducts is also encountered more frequently in patients with right isomerism than in those with pulmonary atresia and an intact ventricular septum.[4,7,8] In our experience, when major aorto-pulmonary collateral arteries have been seen in the setting of right isomerism and pulmonary atresia, the intracardiac anatomy has always been that of an anterior aorta arising from the morphologically right ventricle supported by a muscular infundibulum, in other words the "transposition" rather than the "tetralogy" variant.[4,7,9]

In children with right isomerism and pulmonary atresia, the complexity of pulmonary arterial supply suggests an early morphogenesis of the pulmonary obstruction. Whilst we might also suspect the possible involvement of deficiencies of the neural crest and/or branchial arches, we would then expect the intracardiac anatomy to be that of tetralogy, rather than an anterior aorta supported by a complete muscular infundibulum. From the clinical point of view, the frequency of complex patterns of pulmonary arterial supply associated with the presence of totally anomalous pulmonary venous connection, often obstructed, suggests that accurate pre-operative diagnosis, showing all vascular extracardiac structures, is essential.[1,4,9] In this light, while recognising the significant progress made recently in use of helical computed tomography,[10] we still consider that angiocardiography is necessary.[9]

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Sir,

We thank Piacentini and colleagues for their comments on our article. We agree that aortopulmonary collateral arteries are not so uncommon in association with pulmonary atresia in the setting of Fallot's tetralogy.[1–3] We also recognise that their association with right isomerism is well documented.[4] The association with right isomerism and extracardiac totally anomalous pulmonary venous connection, however, is rare,[5] and this was the echocardiographic diagnosis in our case. Following the successful repair of the anomalous pulmonary venous connection, we did not expect high pulmonary blood flow. Further investigation in the form of catheterisation and angiography revealed redundant aortopulmonary collateral arteries in presence of pulmonary arteries of reasonable size. The successful occlusion of these arteries with coils optimised the flow of blood to the lungs, and helped us wean the patient successfully from ventilation. Our case warranted publication based on the interesting temporal profile of discovery of this rare association. We ourselves also emphasized the indications for cardiac catheterisation and angiography.

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