

Conotruncal (outflow tract) defects: is the ventricular septal defect always the same?

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Background: The ventricular septal defect (VSD) in conotruncal (outflow tract) defects opens into the right ventricle between the limbs of the septal band. However, the terminology of the VSD is ambiguous, particularly in tetralogy of Fallot (TOF): the conoventricular VSD, due to the malalignment between the outlet septum and the muscular septum, is also described as perimembranous because of the presence of a fibrous continuity between the aortic and tricuspid valvar leaflets.

Aim of the study: To analyze the anatomy of the VSD in hearts with conotruncal (outflow tract) defects, in order to solve this nomenclatural ambiguity.

Material and methods: We reviewed 196 heart specimens from the anatomic collection of the French Center of Reference for Complex Congenital Heart Defects: 63 TOF, 54 TOF with pulmonary atresia (TOF-PA), 54 common arterial trunk (CAT), 16 interrupted aortic arch type B (IAA-B) and 19 malalignment VSD without outflow tract obstruction (MVSD). Special attention was paid to the borders of the VSD, viewed from the right ventricular side.

Results: The VSD was conoventricular, located between the 2 limbs of the septal band, in all hearts. There was a fibrous continuity between the tricuspid and aortic valve in 74% of MVSD, 67% of TOF, 24% of TOF-PA, 2% of CAT, 0% of IAA-B ($p < 0.005$). The aortic valve was continuous with the anterior tricuspid leaflet, but not with the septal leaflet, contrary to perimembranous VSD.

Conclusion: All conotruncal (outflow tract) defects share the same VSD, conoventricular in type. However, there are some differences between these defects regarding the inferior rim of the VSD. The continuity of the aortic valve with the anterior tricuspid leaflet (and not the septal leaflet) indicates that this continuity may be a consequence of the deviation of the ventriculo-infundibular fold, along with its outlet septal component, rather than a perimembranous extension of the VSD. Finally, these differences suggest that conotruncal (outflow tract) defects should be considered as an anatomic continuum rather than distinct physiological phenotypes.