Nikaidoh Procedure for Double-Outlet Right Ventricle with a Subpulmonary Ventricular Septal Defect and Pulmonary Stenosis

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Double-outlet right ventricle with a subpulmonary ventricular septal defect (VSD) and pulmonary stenosis (PS) is an infrequent congenital heart defect. The surgical management of this lesion remains challenging, commonly requiring palliation before proceeding with the definitive surgical repair.

The Nikaidoh procedure is one of the established surgical techniques for the management of this complex CHD. It consists of harvesting the aortic root from the right ventricle (with or without one or both coronary arteries attached), reliving the LVOTO (dividing the outlet septum and excising the pulmonary valve), reconstructing the LVOT (with the posteriorly translocated aortic root and the VSD patch) and the right ventricular outflow tract (RVOT) with a conduit or a pericardial patch. Although technically challenging, this approach is felt to result in a “more normal” anatomic repair.

Several modifications to the original technique have been described, but the main technical point remains the repositioning of the native aortic root closer to the left ventricular cavity, avoiding the creation of a long tortuous intraventricular tunnel. This
technique appears to prevent the development of LVOTO, which is a frequent complication of the Rastelli repair.

Anatomical studies and clinical experience has helped identify a subset of patients that appear to be better managed with a Nikaidoh procedure. These include patients with the following findings:

- Inlet type or more apically located VSDs
- Hypoplastic RVs
- Straddling AVV

The presence of a straddling tricuspid valve has been reported as a risk factor for early and late death with the Rastelli repair.

Aortic root dilatation and/or aortic valve insufficiency has been observed in some patients after the Nikaidoh procedure. The etiology of these findings are still unclear, but they are likely similar to those affecting the pulmonary autograft after a Ross procedure. In our experience aortic valve insufficiency has only developed in patients in which both coronary arteries required reimplantation.

Although the overall experience with aortic translocation is limited, the early and midterm results are encouraging. Certainly, more long-term data is needed.


