Operative Techniques in
Thoracic and
Cardiovascular Surgery

Ligation of Right Ventricle to Coronary Artery Connections to Allow a Two-Ventricle Repair Track in Patients with Pulmonary Atresia and Intact Ventricular Septum

John E. Foker, MD, PhD,* James M. Berry, RDMS,† Brian A. Harvey, BA,* and Lee A. Pyles, MD†

The complex spectrum of lesions within the diagnosis of pulmonary atresia with intact ventricular septum (PAIVS) continues to pose difficulties in effective treatment. Most of these infants end up with a single ventricle repair (SVR) or more palliative endpoint and mortality remains high.[1] A 2-ventricle repair (2VR) is more desirable, but there are several obstacles to it, beginning with the frequent severe hypoplasia of the right ventricle (RV) and tricuspid valve (TV). In addition, large connections between the RV and the coronary arteries (RV-CACs) are common and are considered to prevent a 2VR.

The RV-CACs are found in about 30 to 40% of all PAIVS infants but are much more frequent with severe RV hypoplasia. When significant RV-CACs are present, decompression of the RV on bypass or by relieving the outflow obstruction may produce a severe, and even lethal, myocardial steal.[2,3] Consequently, significant RV-CACs are commonly thought to preclude any attempt to achieve a 2VR.[2]

The right heart hypoplasia of PAIVS, however, is developmental rather than primarily genetic in origin and, therefore, catch-up growth resulting in a 2VR should be possible with the correct growth signals.[4,5] The considerable long-term advantages of 2-ventricle physiology have led us to pursue this approach in essentially all PAIVS infants, despite the obstacles. Because significant RV-CACs make bypass unsafe, it was first necessary to develop techniques to take down these connections off bypass.

The significance of the RV-CACs is complicated by the associated coronary artery pathologic condition. The RV-CACs vary in size, number, and location and because the coronary artery (CA) flow is often largely retrograde, under high pressure and with very hypoxic blood, secondary stenotic myointimal lesions and myocardial damage are also frequently found.[6,7] The obstructive lesions found in the arteries themselves complicate the evaluation of the RV-CACs because they may increase the consequences of a myocardial steal. The diagnostic studies may also suggest the circulation is dependent on the RV for flow (Fig. 1). As a result, any combination of significant RV and TV hypoplasia together with RV-CACs has effectively prevented a 2VR track. The advantages of a 2VR over a SVR, however, justified our pursuing this approach despite the considerable obstacles.[8] The techniques developed for ligation of the RV-CACs have been successful and we recently reported our experience with this approach.[9]

Further justification comes from recent studies that have shown the RV-CACs do not reliably involute and the intimal lesions and myocardial injury may progress.[7] The inability to decompress the hypertensive RVs, moreover, leads to increased LV dysfunction in SVRs.[10] Consequently, ligation of the connections and RV decompression seem desirable in all PAIVS patients even if a 2VR is not achieved.

The location and ligation of significant RV-CACs require close cooperation among the cardiology, echocardiographic, and surgical teams during the various stages of evaluation and treatment. The only situation that definitely precludes RV decompression and placement on a 2VR track is the developmental absence of a connection between the aorta and the main CAs. The presence of aorto-coronary artery continuity, therefore, must be established before addressing the RV-CACs. When shown by angiogram (Fig. 1), it is most clear but the finding of a diastolic antegrade flow signal, however brief, also proves continuity.[11] When no continuity is present, the patient is best treated by urgent transplantation.

With aorto-coronary continuity established, the coronary artery anatomy must be defined and the RV-CACs located (Fig. 2). The RV-CACs and significant intimal lesions within the CAs may suggest a RV-dependent circulation. We found it difficult to judge RV dependency preoperatively, however,

*Division of Cardiothoracic Surgery, University of Minnesota, Minneapolis, Minnesota.
†Division of Pediatric Cardiology, University of Minnesota, Minneapolis, Minnesota.
This work was supported in part by the Robert and Sharon Kaster Endowment for Pediatric Cardiac Surgical Science. Dr. Foker reports equity ownership in Bioenergy, Inc. Dr. Harvey and Dr. Pyler have no commercial interests to disclose.
Address reprint requests to John E. Foker, MD, PhD, Robert and Sharon Kaster Professor of Surgery, Division of Cardiothoracic Surgery, University of Minnesota, MMC 495, 420 Delaware St. SE, Minneapolis, MN 55455. E-mail: foker001@umn.edu

1522-2942/$-see front matter
doi:10.1053/j.optechstcvs.2010.07.003

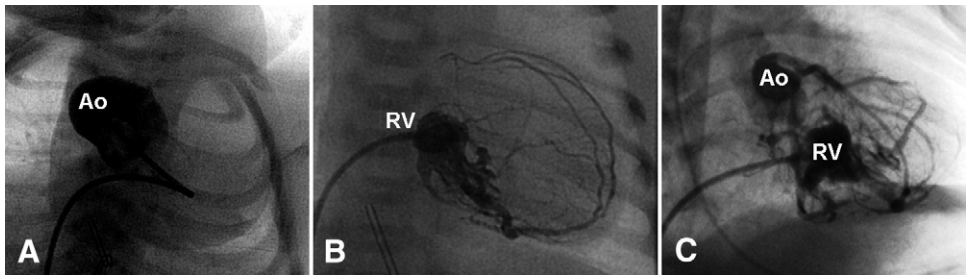

Figure 1 The first views. The diagnosis of PAIVS is typically by fetal ultrasound and at that time an estimation of the degree of right heart hypoplasia can be made. Although RV-CACs are not often seen in utero, they are commonly present with severe right-heart hypoplasia and their presence can often be inferred. Ligation will require fairly precise localization, however, and more definitive studies are required postnatally. If the postnatal transthoracic echocardiogram confirms the presence of RV-CACs, angiograms will be very helpful. During the catheterization studies, pulmonary blood flow will be maintained by prostaglandin E_1 infusion.

(A) An injection of the proximal aorta (Ao), which fails to reveal the coronary arteries, may occur because their perfusion is retrograde through the RV-CACs from the hypertensive RV. An image like this may be daunting when combined with the accompanying RV gram.

(B) The RV injection often best reveals the coronary arteries and begins to define their anatomy and the RV-CACs present. In this case, an RV-dependent CA circulation was also suggested.

(C) Because of the preceding aortogram and RV gram, it must be established that the developmental connection between aorta and the coronary arteries has been made. This is most easily done when the aorta fills from the RV gram as shown here from another patient or when the aortogram fills the CAs. Otherwise it will have to be determined by echo studies, which look for an often brief antegrade flow in the main CAs. Ao = aorta; RV = right ventricle.

and in practice it appears to be infrequent.[9] The ligation of the RV-CACs eliminates this component and also provides information about the significance of the intimal lesions. Because the ligations of the RV-CACs are readily reversible, the question of RV dependency need not be answered preoperatively and can be determined at operation. In our report of 71 ligations in 19 patients, no ligations were removed.[9]

The preoperative assessment using angiography and echocardiography reveals the likely site of significant connections but, for those that are not readily found, intraoperative echo techniques must locate them. As expected, the operative location and ligation of the RV-CACs pose some difficulties. The methods used have varied with the anatomy of the connections and the technical details are a major component of this report.