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.
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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

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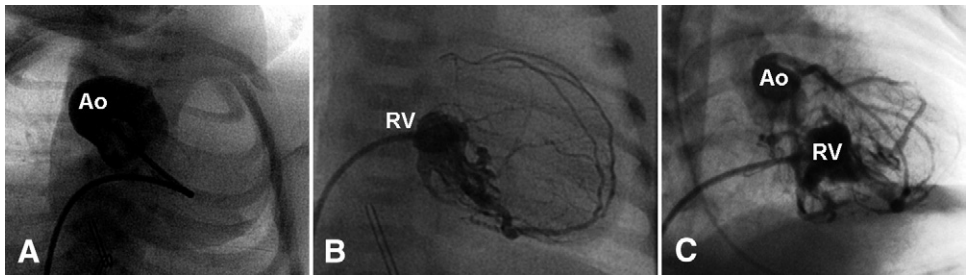


Figure 1 The first views. The diagnosis of PAIVS is typically by fetal ultrasound and at that time an estimation of the degree of right heart hypoplasia can be made. Although RV-CACs are not often seen in utero, they are commonly present with severe right-heart hypoplasia and their presence can often 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.

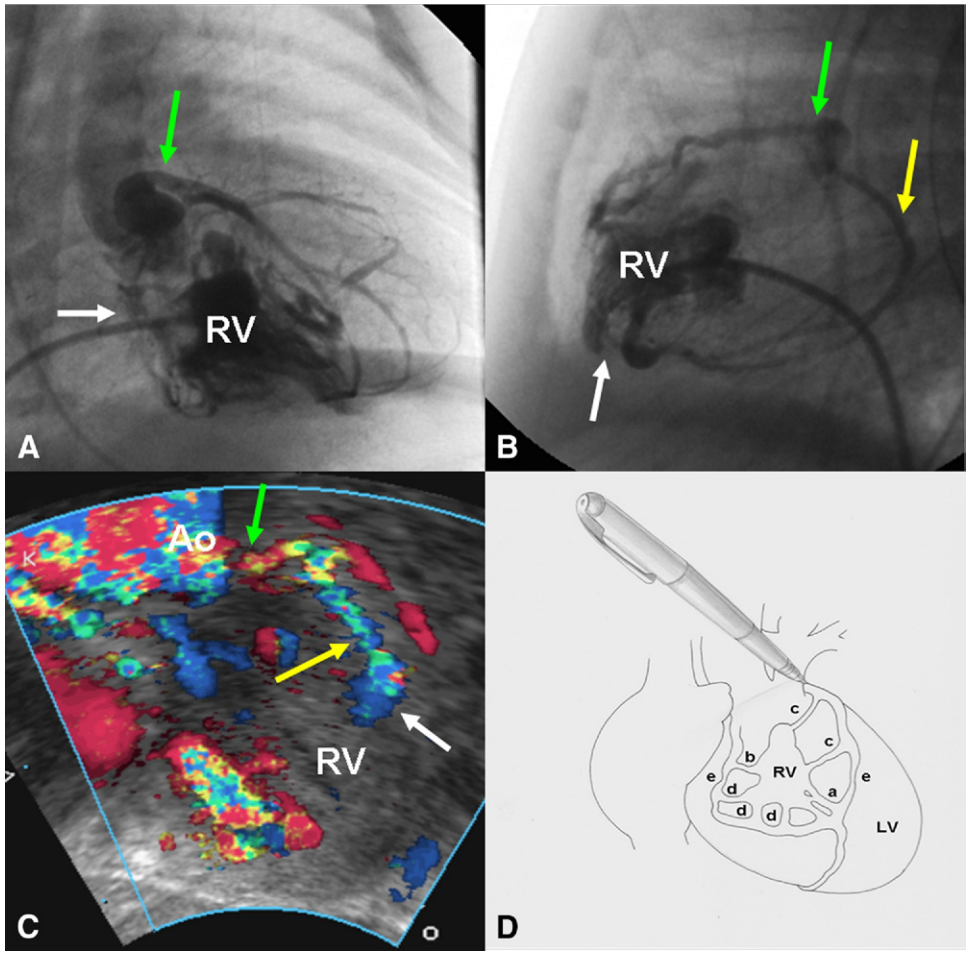


Figure 2 Preoperative assessment. Ligation of the RV-CACs requires a detailed preoperative assessment so the connections can be found and ligated off bypass. Moreover, the significant intraluminal lesions and obstructions that may affect the consequences of RV-CAC ligation should be determined. Together, these 2 types of lesions may have profound consequences for myocardial perfusion and typically have already produced significant damage in these newborns.[6]

To generate a preoperative diagram of the RV-CACs that will be helpful in the operating room, more detailed right ventricular angiograms and echocardiograms are needed. The suprasystemic RV pressures produce more retrograde than antegrade coronary artery flow and the RV studies commonly provide the best filling of the coronary arteries. The right anterior oblique and lateral angiographic projections are the most helpful views to define the connections.

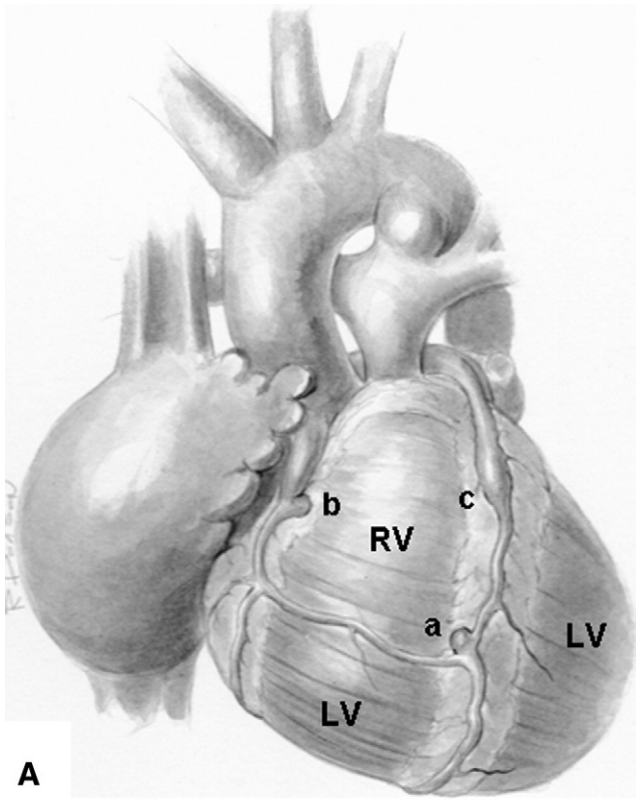
(A) The left main (green arrow) and left anterior descending (LAD) coronary arteries as well as the right coronary artery (RCA) (white arrow) are demonstrated in this view (right anterior oblique with caudal angulation). Significant RV-CACs are visible at the RV apex and along the posterior descending artery. Continuity with the aorta is also established. (See Video 1; supplementary videos are available online at http://www.optechtcs.com.)

(B) The LAD (green arrow) and left circumflex coronary artery (yellow arrow) are shown in this lateral projection of RV angiogram. The RV-CACs off the RCA system out to the apex are also further delineated. For these figures only single frames can be presented; however, playing the angiograms back and forth provides considerable additional information and better reveals the number, location, and size of the RV-CACs (Video 2).

(C) The transthoracic echo evaluations do not provide as precise anatomic detail; nevertheless, because of the different angles of visualization and dynamic nature, additional connections may be revealed and occasionally a better idea of their relative size may be gained. The left main (green arrow) and LAD arteries (yellow arrow) are shown. The RV is contracted and the LAD flow is retrograde with systole. A significant RV-CAC (white arrow) is shown by a wider blue jet in the area of the RV apex (Video 3).

(D) After thorough preoperative evaluation, a sketch can be made that diagrams the location of significant RV-CACs and the sites of apparent stenoses. This diagram was generated from the 2 angiographic projections and the transthoracic echo study. The studies revealed a large RV-CAC at the apex (a), which by 1 view, may have more than 1 channel from the RV to the CAs. A somewhat larger ligation may be needed to incorporate them and eliminate this connection. The RV-CAC from high on the RCA appeared to be smaller, but nevertheless, significant (b). The connections from both the left anterior descending (LAD) coronary artery and those from the RCA are relatively straightforward and apparently single (c). The RV-CACs at the angle of the right coronary artery also appeared to have several channels, which the surgeon must be aware of at the time of ligation (d). The stenoses located (e) should alert the team that ligation of connections more distally might produce a WMA. This type of diagram is helpful both from the analysis that goes into it and for the road map it provides. Ao = aorta; LV = left ventricle; RV = right ventricle.

Operative Technique



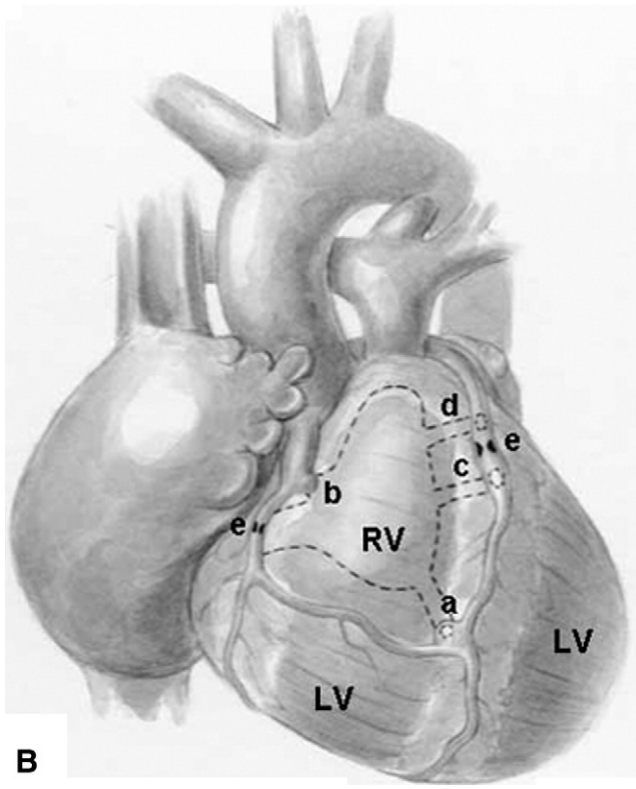


Figure 3 (A) After opening the pericardium of a patient with PAIVS, the unusual surface anatomy is apparent. The left anterior descending artery (LAD), which normally is just around the corner of the left heart border, is plainly seen coursing down the anterior surface of the heart. Similarly, the posterior descending portion of the RCA is visible well up from the lower ventricular border. These coronary arteries outline what is clearly a hypoplastic RV.

When RV-CACs are present, the hypoplastic RVs are typically hypertensive and the coronary artery perfusion is retrograde with very hypoxic blood. Postmortem studies have indicated this leads to significant myocardial damage.[6] As a result, the surface of the myocardium is often blotchy and yellowish in appearance and irritable. Although we have had no sustained dysrhythmias from the maneuvers used to ligate the RV-CACs, they remain a possibility. If a significant dysrhythmia occurs, rescue by bypass will place the myocardium at increased risk from a steal and should be avoided until the significant RV-CACs are ligated. The placement of cannulation site sutures and looping the PDA before the RV-CAC ligations will facilitate rapid conversion to support if needed.

This figure illustrates the different epicardial clues that help locate the larger connections that have been identified preoperatively. A lateral, dome-shaped protuberance indicates the presence of a large connection. This feature is most clearly seen when it occurs at the junction of the LAD, RCA, and circumflex arteries (a). Once the significance of this configuration is understood, it will be useful to identify similar findings off the LAD artery or RCA (b). Another clue to a significant connection is a relatively sharp drop in diameter of the CA (c). This change in configuration signals a connection arising posteriorly from the CA, which dives directly to the RV rather than coming off laterally.

(B) With the likely sites of the RV-CACs determined from the preoperative studies and the clues to their exact location from the surface features of the coronary arteries (a), the surgeon will be able to predict the internal anatomy of the connections and what must be considered for the ligations. This figure illustrates the several types of surface clues to the presence of RV-CACs as well as the predicted internal anatomy of the connections and the hypoplastic RV. During this time the TE echo study done at the beginning of the case should reinforce these conclusions, and possibly, identify additional, unsuspected RV-CACs. The TE echo assessment will also be helpful as the surgeon proceeds with the ligations. LV = left ventricle; RV = right ventricle.

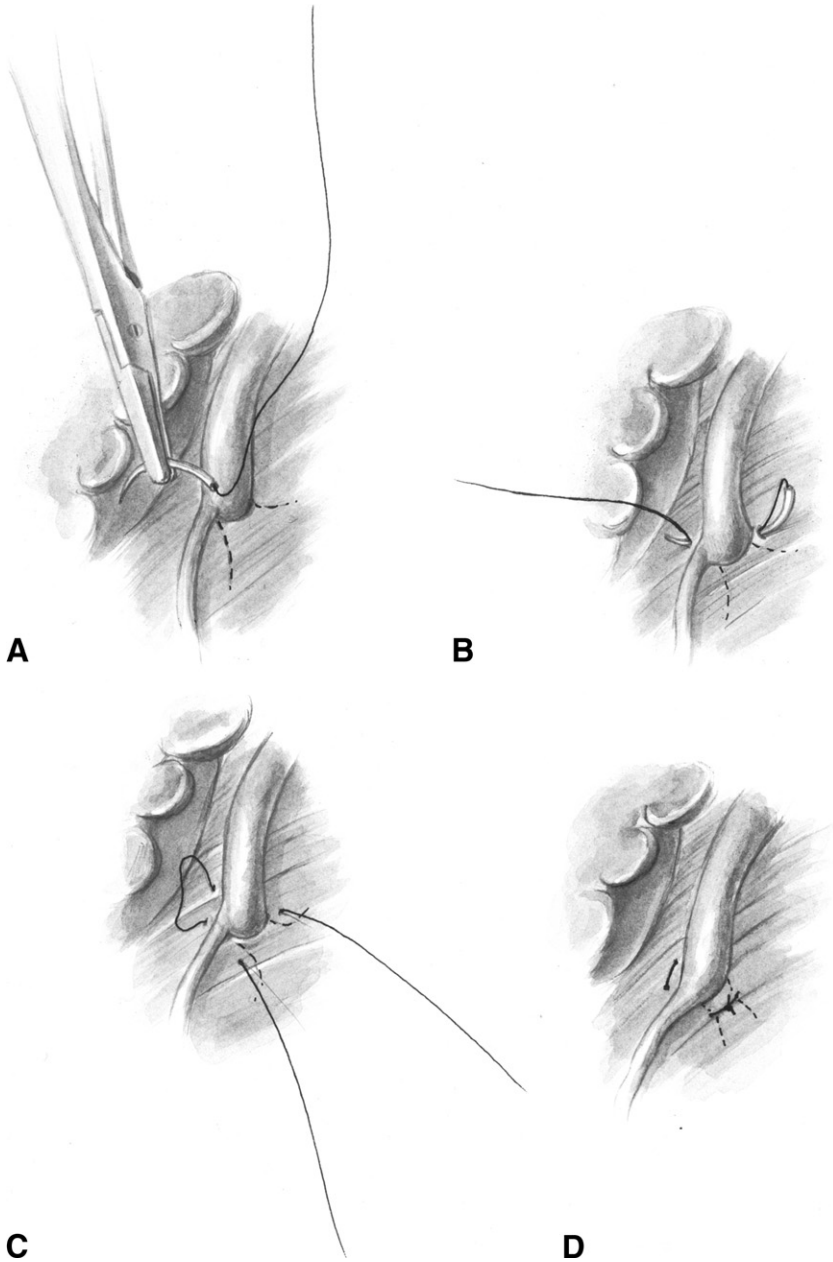


Figure 4 Techniques used in locating and ligating the RV-CACs. The methods of locating and ligating the RV-CACs are depicted in this series of drawings. The series of ligations we have reported were done with either 7-0 or 6-0 Prolene sutures on the smallest gauge needle.[9] Although one could do the ligations by placing the needlepoint forward, if the needle is backed through, it can be gently worked through the tissues producing less laceration and bleeding. Very little bleeding was encountered and in no case did the suture placement produce any discernible damage. As depicted, the needle is passed underneath the epicardial coronary artery on both sides of the connection as a horizontal mattress suture. The needle can be placed closer to the coronary artery than depicted in the illustration, although because this is a horizontal mattress stitch, it should not be so close as to crimp the coronary artery with tying.

The technique used to ligate the RV-CACs with characteristic surface appearances is presented in A-E. For a connection originating as a lateral protuberance, the needle is backed underneath the coronary artery on each side and tied down (A-D). The echo team should see, at the least, a decrease in retrograde coronary artery flow in the vicinity. The connections themselves are not always easily visualized by TE echo and the change in direction of coronary artery flow may be the best method of determining that ligation has taken place. The echo team should also look for WMAs, particularly in the apical-septal area, which is perfused last by coronary artery flow.

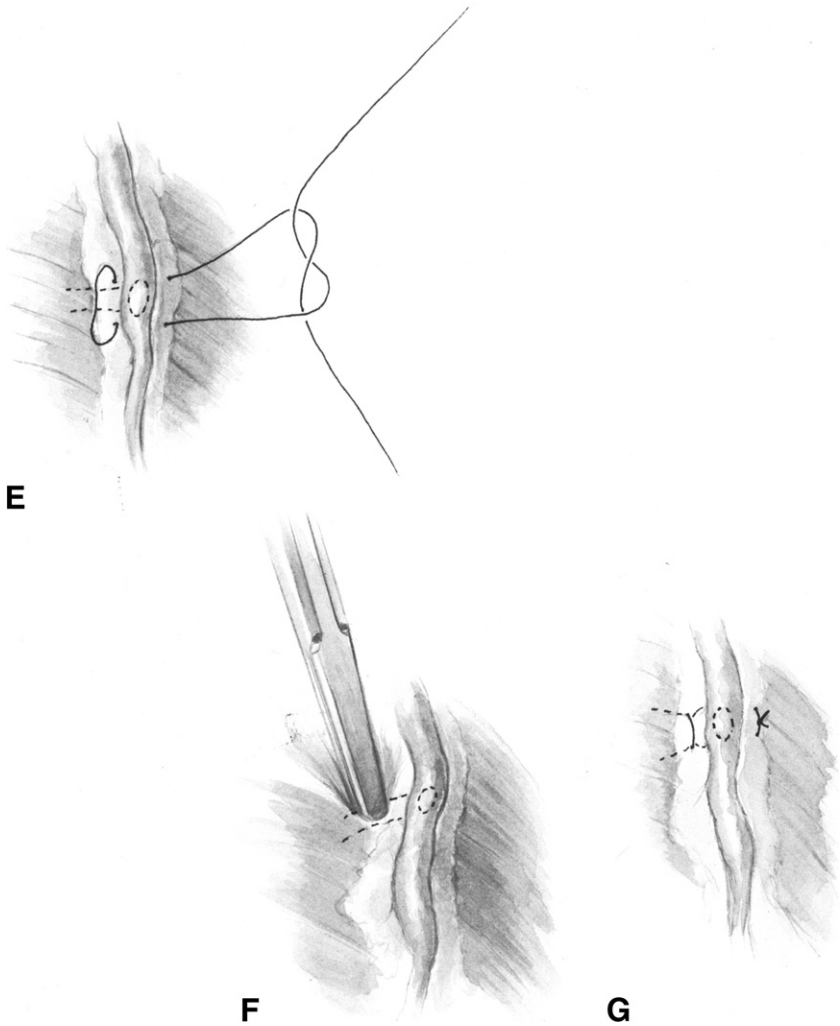


Figure 4 (*Continued*) (E) A second clue to the presence of a connection is a discrete decrease in the diameter of the coronary artery. This location should also correspond to the preoperative identification of a significant connection and confirmation can be achieved by pressure with a blunt pick-up or similar instrument in the area with a resulting change in the direction of coronary artery flow or, in some cases, interruption in flow within the connection itself can be seen.

(F, G) Sometimes, the configuration of the coronary artery does not provide a surface clue to the location of the connection, which has been identified preoperatively. The location may be identified by using gentle pressure with a blunt pickup along the course of the coronary artery (F). When the connection is compressed, the change in signal can be identified by the echo team. Ligation can then be performed and the connection eliminated (G).

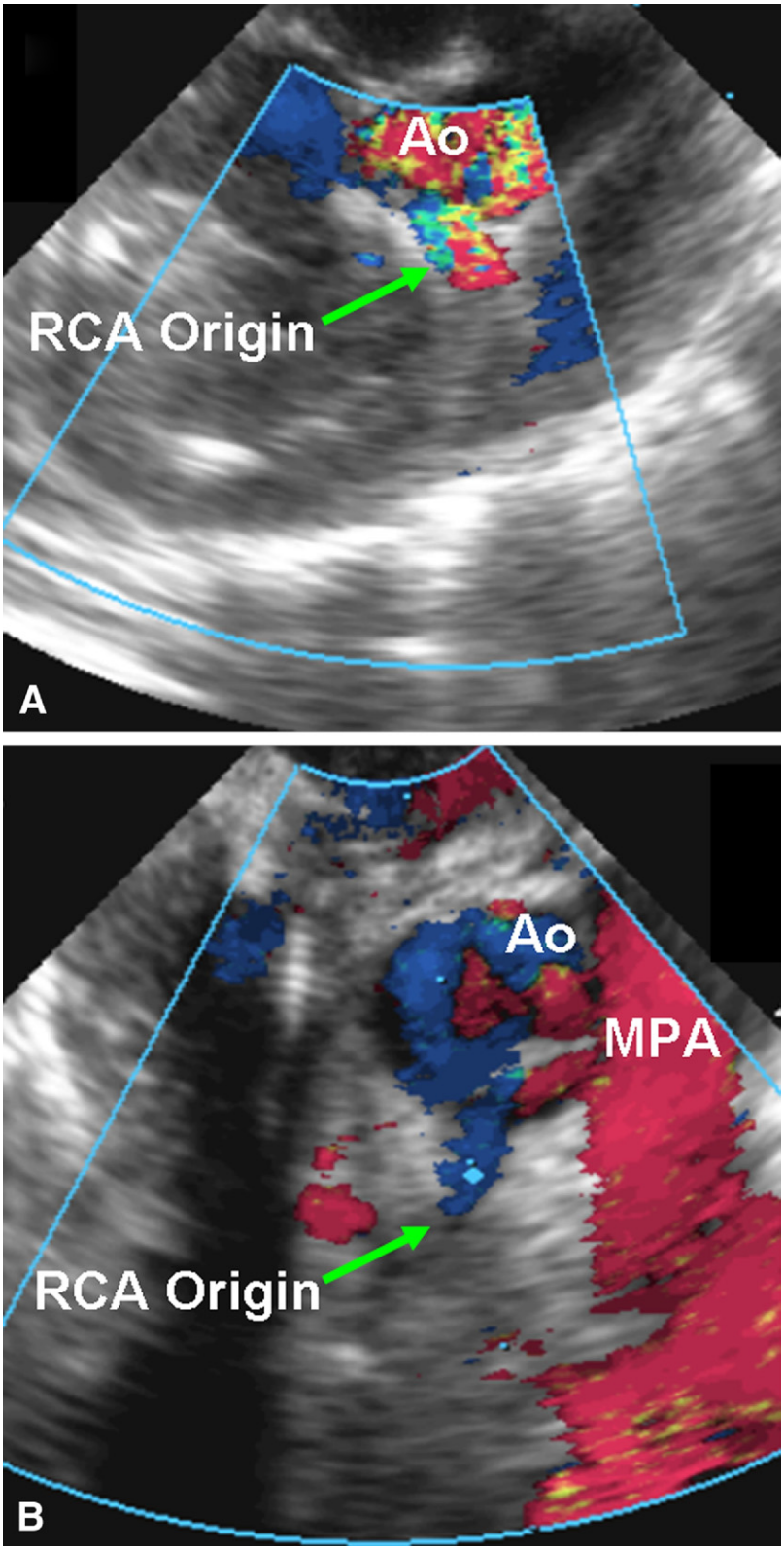


Figure 5 Coronary artery flow signals are used initially to establish continuity with the aorta and, later, to assess the effects of RV-CAC ligation. In this case, flow between the aortic root and the right coronary artery (RCA) is shown before and after ligation of a more distal RV-CAC. (A) Transesophageal echocardiography revealed retrograde flow from the RCA into the aortic root, which demonstrated both aortic-coronary continuity and the presence of higher pressure in the connection with retrograde flow into the aorta (Video 4 and Video 5). (B) After ligation, only antegrade flow into the RCA from the aortic root was seen, indicating elimination of the connection (Video 6). Ao = aorta; MPA = main pulmonary artery; RCA = right coronary artery.

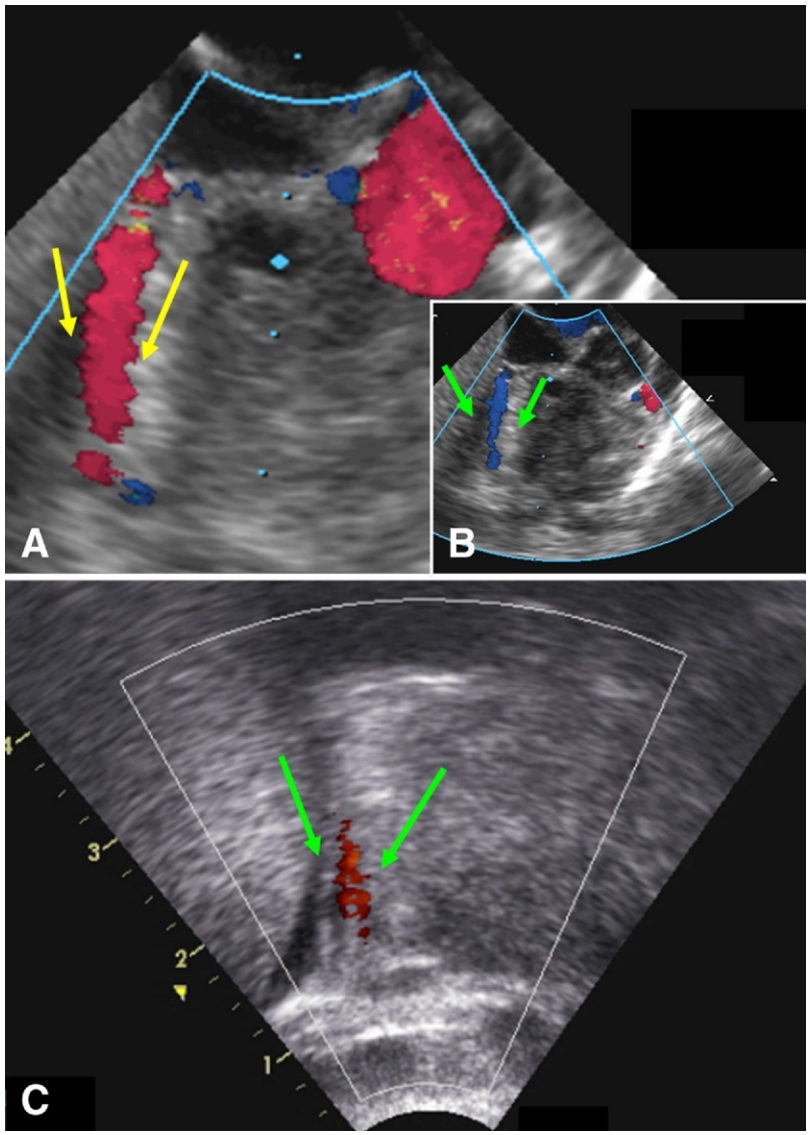


Figure 6 Assessing RV-CAC ligation by TE echo. The effect of ligation of RV-CACs on coronary artery flow can be determined using TE (Video 7). (A) Initially, flow in the posterior descending coronary artery (PDCA) is predominantly retrograde in systole (yellow arrows) from a large apical RV-CAC. (Image from the gastro-esophageal junction at 93° angulation.) (B) (insert) The same study shows brief antegrade flow in diastole in the posterior descending (PD) CA (signal between green arrows), which, in the absence of a more proximal connection, confirms continuity with the aorta. (C) After ligation, PDCA flow is entirely antegrade (normalized) in this different view—a modified apical 2-chamber projection. (The ventricular septum is en face and the PDCA is oriented opposite from the image in (A).)

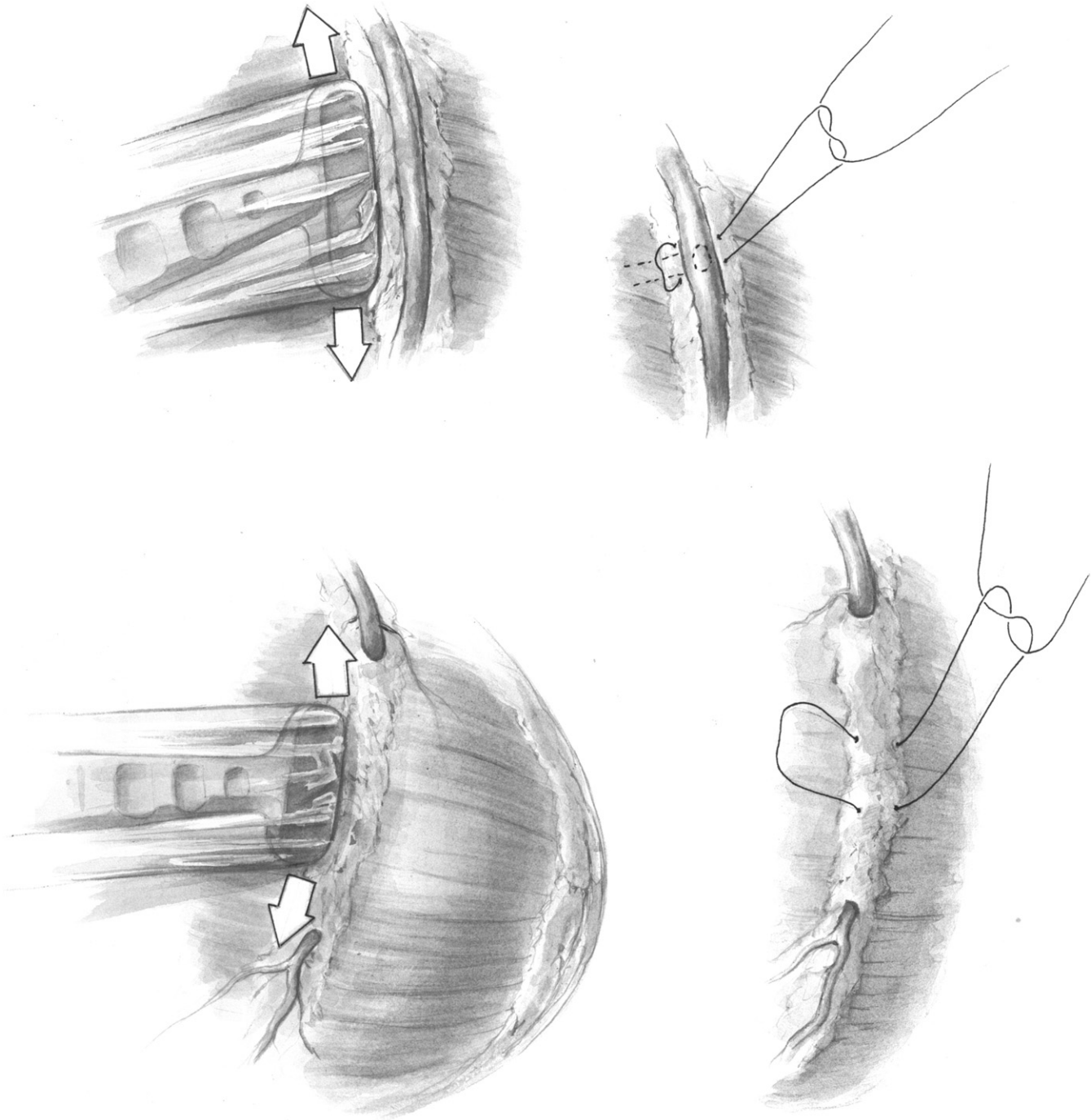


Figure 7 On occasion, all of the surface clues and techniques presented are not sufficient to reveal the location of the connection and surface echo must be employed. By moving a high-definition (15 MHz linear array) hockey stick probe along the course of the coronary artery, the location of the connection may be revealed and ligation performed.

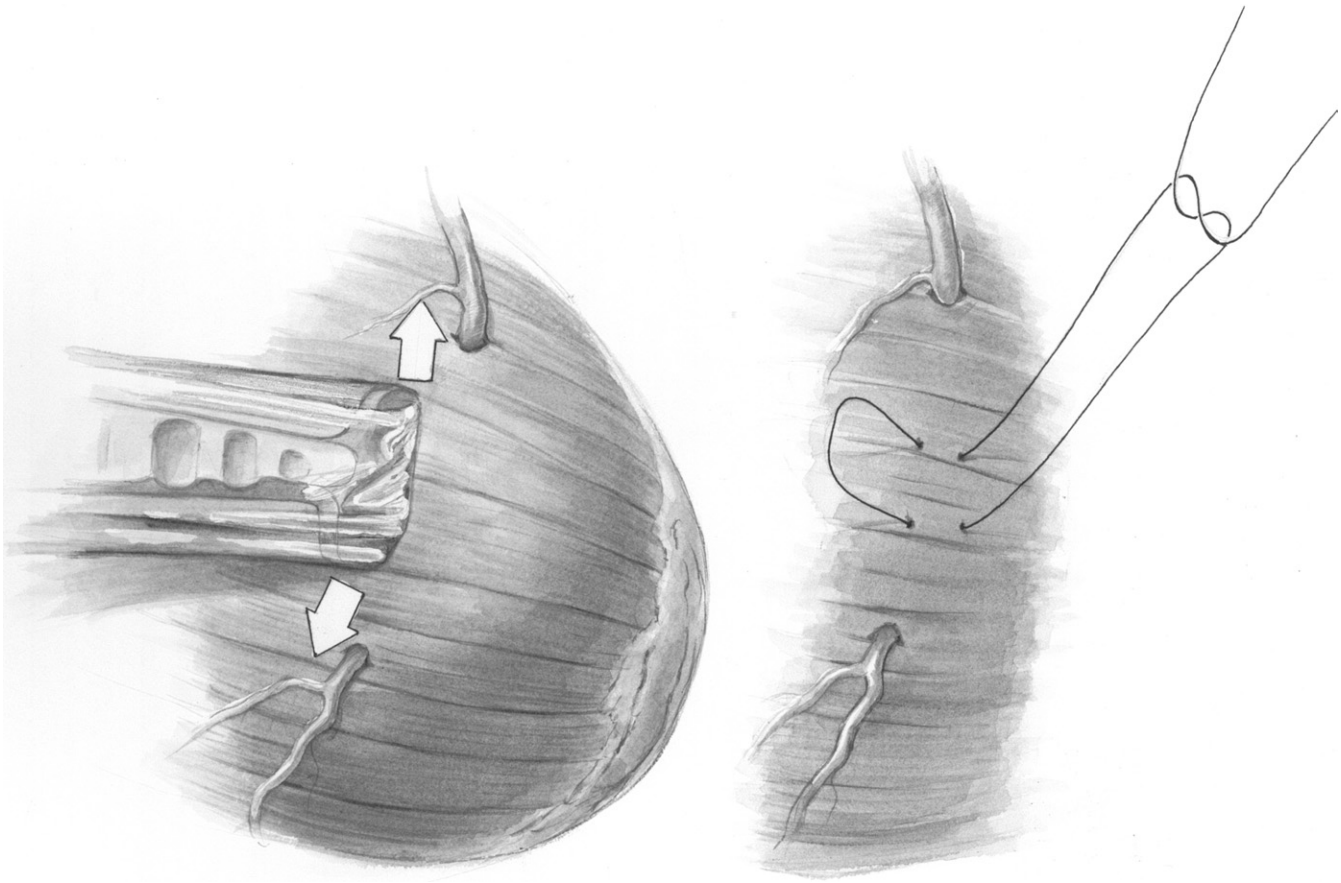


Figure 8 For the occasional case, where much of the coronary artery is intramyocardial, locating the connection can be difficult. We have found the high definition hockey stick probe also to be useful in locating these connections. The probe is moved back and forth along the presumed line of the coronary artery to localize the signal and, therefore, the site of the connection. Once determined, the site is confirmed by pressure with a blunt instrument as before. The ligation then is done in a similar fashion and tying down the suture should provide a confirming change in the signal.

With a little experience by both the surgeon and the echo colleagues, the ligation of the significant connections has proven to be surprisingly straightforward for the team.

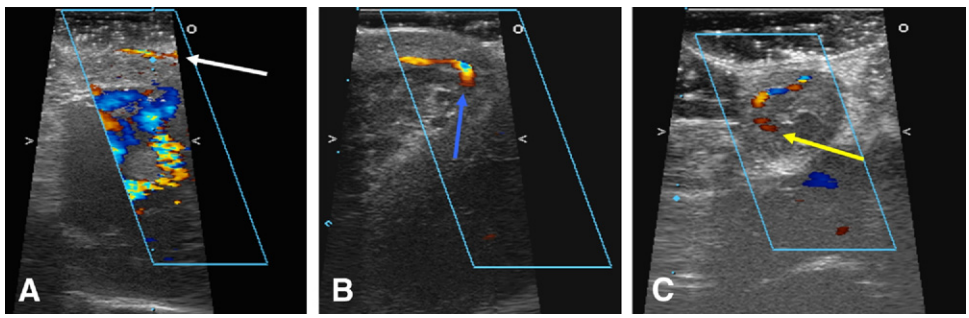
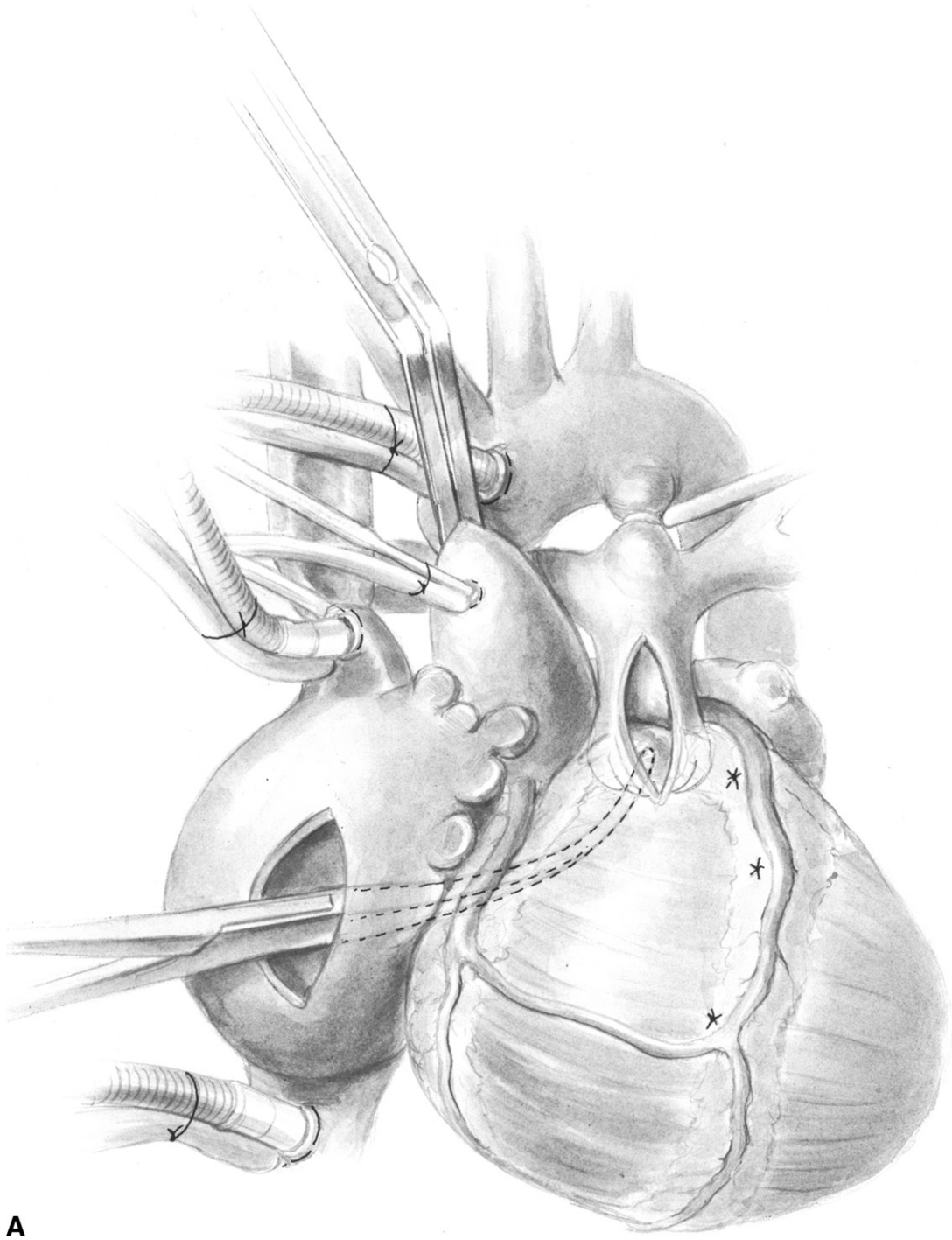


Figure 9 The surface echo studies using a 15 MHz hockey stick transducer in a sterile sleeve used to locate a connection that is not apparent from the surface anatomy of the coronary artery (Video 8). (A) The coronary artery (white arrow) is defined in this image from the RV surface. (B) By moving the transducer back and forth near the coronary artery, the RV-CAC signal is located (blue arrow) as it connects to the RV chamber. (C) Another RV-CAC is found coursing from the RV toward the epicardial surface (yellow arrow).

After ligation of the RV-CACs, the repair can be performed on bypass with the aim of inducing right heart growth and achieving a 2VR.[5,12] To maximize growth, the RV outflow tract obstruction must be effectively removed, as even moderate residual obstruction appears to limit regression of the hypertrophy and reversal of the cavitary hypoplasia. The growth signal itself seems to be generated by AV valve flow and, therefore, although right-to-left shunting is necessary initially to maintain cardiac output, the atrial septal defect (ASD) should be mildly restrictive to encourage TV flow.[4,5,12] After the bypass run, the ASD snare is adjusted under transesophageal (TE) echocardiography guidance to about 3 to 4 mm in diameter to encourage TV flow (Figs. 11 and 12) The hypertrophied RV will collapse with decompression further



A

Figure 10 The components of the PAIVS repair to encourage catch-up growth and a 2VR endpoint. After ligation of all significant RV-CACs, the patient is heparinized and placed on cardiopulmonary bypass (A). Aortic cross-clamping with cardioplegic arrest allows the steps to be performed with precision despite the small size of the structures. The PDA, previously looped, is cinched down to improve perfusion and allow opening of the main PA. A right atriotomy incision is made near the presumed site of the ASD and TV. With the right atrium opened, the patent foramen ovale/secundum type atrial septal defect is visualized and the TV is assessed. Even though small, the TV will allow a fine, curved clamp to be passed through it, into the hypoplastic RV cavity. This maneuver makes creating an opening into the body of the RV safer and more effective.

reducing its volume; therefore, stability while growth takes place also requires adequate pulmonary blood flow provided by either maintaining a patent ductus arteriosus or placing a systemic-to-pulmonary artery shunt. Finally, the adequacy of ligation and the search for additional connections are also performed after bypass.

A number of anatomic situations may be encountered in these complex patients but with the cooperation outlined between surgeon and echo team, the location and ligation of the connections has proven surprisingly straightforward. Again, a very important consideration is that each ligation is readily reversible. If ligation produces a significant wall mo-

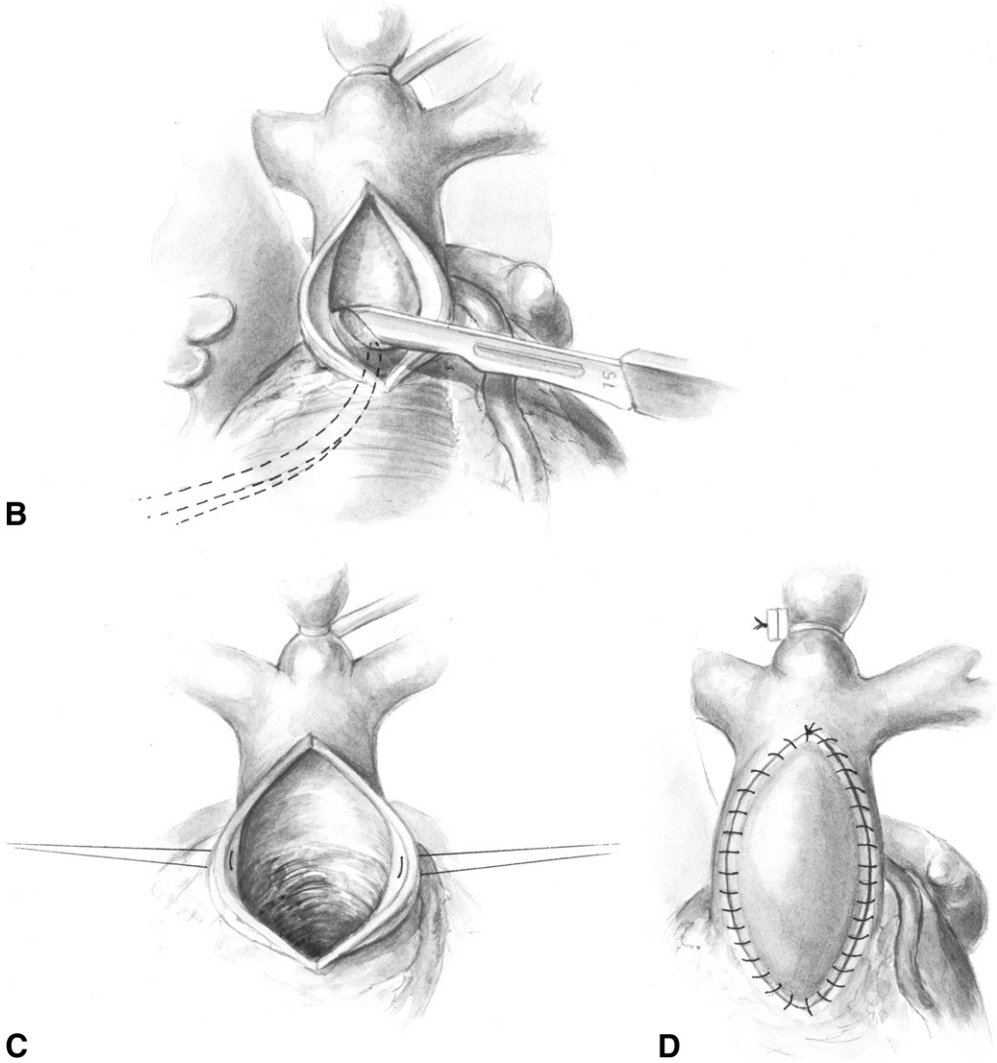


Figure 10 *(Continued)* The main PA is opened vertically, exposing the atretic PV. At the more severe end of the spectrum, what would be the area of the infundibulum is an essentially solid core of muscle. By cutting on the tip of the clamp, the small RV cavity can be entered and continuity established so the complete relief of obstruction can be performed safely (B).

The RV muscle is cored out so that, at most, only minimal outflow obstruction remains (C). This can be performed without damaging the papillary muscles within the hypoplastic RV cavity.

With a nonobstructive passageway into the body of the RV established, the outflow tract is patched open with autologous pericardium. The RVOT will reliably grow if the obstruction is relieved initially and flow is present across the TV. The RVOT patch, therefore, is kept modest in size, about 1.5 cm in widest diameter, to keep the unfavorable mechanical effects of the patch on the RV as minimal as possible (D).

With decompression, the hypertrophied, poorly compliant RV collapses further and pulmonary blood flow would be very inadequate by this route. At the very least, ductal patency must be maintained with prostaglandin E_1 infusion. Two factors will govern right-sided forward flow: RV compliance and RV/TV size. If the RV volume is generally adequate in size ($z \geq -4.0$), then 7 to 10 days of PGE_1 infusion may be adequate to allow sufficient improvement in RV compliance to have adequate forward pulmonary flow. Because the PDA may become quite large with continuous PGE_1 infusion, the PDA size can be controlled with a 4-0 Prolene as shown. The retrograde abdominal aortic flow reversal should only be mild to moderate to prevent cardiac failure. This adjustment can be made using TE echo assessment and the snare clipped to fix it. For a newborn, the diastolic pressure should be >30 mmHg.

For a smaller RV, a period of growth will be necessary and a more practical solution will be to place a shunt between the aorta and RPA.

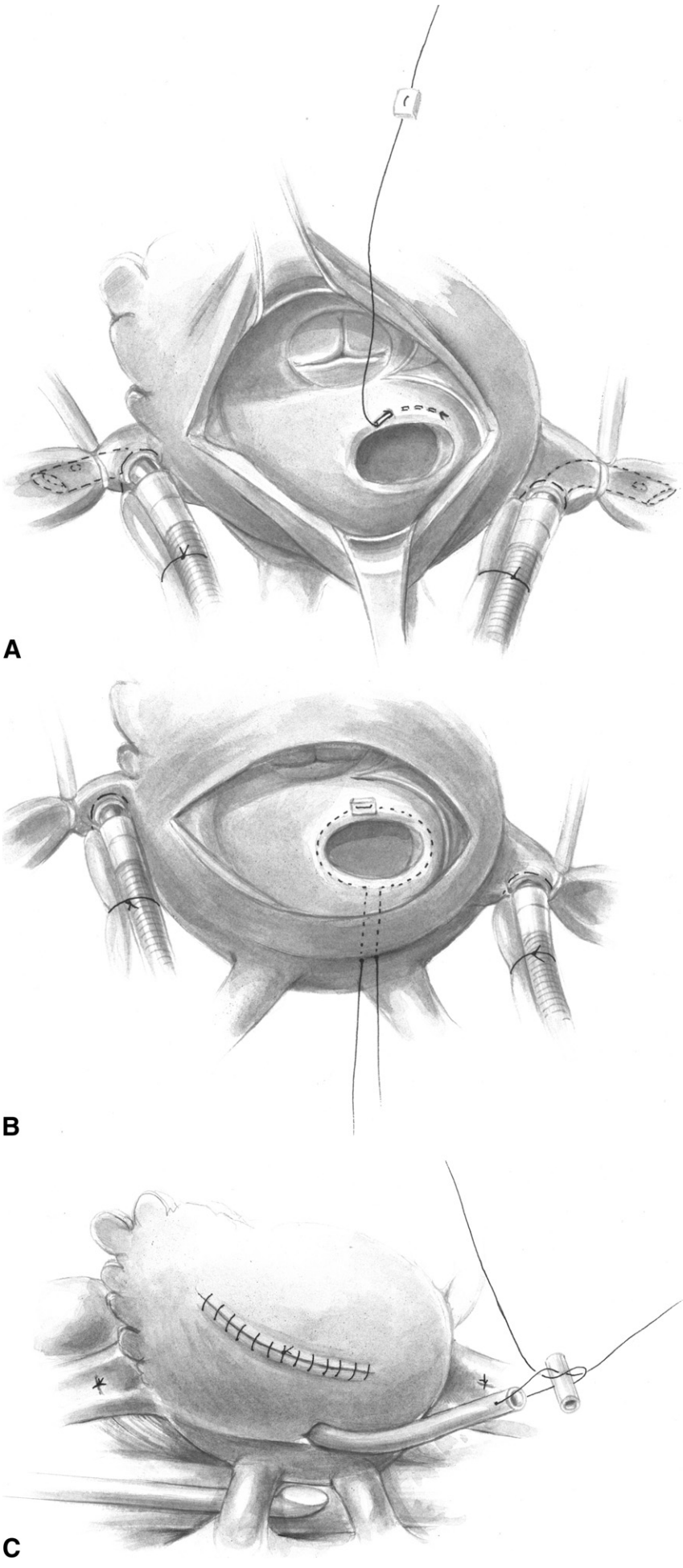


Figure 11 Growth of the right-sided structures depends on flow across the tricuspid valve and, therefore, this flow must be encouraged otherwise these structures will remain hypoplastic. If the septum primum is deficient and does not reach the superior margin of the ASD, the defect may be too large and nonrestrictive. If the septum primum flap is longer and reaches to the superior margin of the defect, although eventual closure should take place with growth of the RV/TV and a fall in right-sided pressures, a controlled size is desirable and the defect is usually snared.

(A) The defect is snared by placing a pledgetted 4-0 Prolene suture beginning at the medial aspect. The suture is placed virtually entirely within the atrial septum itself and far enough away from the edge to insure holding power for the snare. To accomplish this, the needle passes through as much tissue as conveniently possible and then brought out to the surface of the septum.

(B) The needle is regrasped and continues its journey within the septum until it can be brought out the intra-atrial groove as shown. Along the lower margin of the defect, the septum primum is reefed up until the atrial septum itself is again reached.

The goal of placing the snare is to create a single channel through the intra-atrial septum so that its size can be effectively regulated. If there are large gaps in the suturing, the folds and channels will appear as the snare is tightened and it will be difficult to accurately adjust the defect.

(C) The snare is brought out through a piece of silastic tubing with the sutures placed through the wall near the end to provide stability when tying over another short piece of plastic tubing. Notching of the end of the longer tube will stabilize the short piece of tubing.

The adjustment of the ASD size occurs later when off bypass and under echo guidance.

As experience has also shown with fenestrations for the Fontan procedure, a small hole will allow significant right-to-left shunting. A well-defined single defect will allow the shunting to be adjusted off bypass. Later, if desirable, closure can be done by either tightening the snare or placing an ASD closure device.

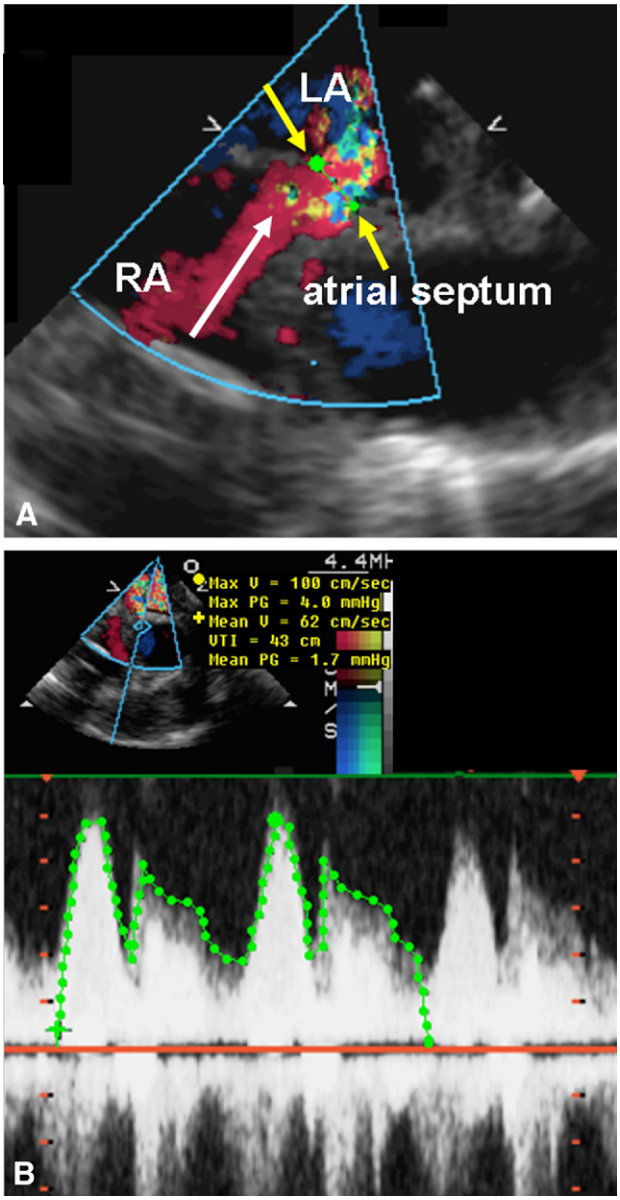


Figure 12 Flow across the ASD is adjusted by echo guidance of the snare off bypass (Video 9).

If the PDA remains open to provide pulmonary blood flow, the PDA snare can also be adjusted under echo guidance to minimize failure. The PDA loop is adjusted to avoid excessive runoff (a mild-moderate abdominal aortic flow reversal is desirable) and to prevent further enlargement with continued PGE_1 infusion.

To encourage TV flow and right-sided growth, the ASD must be mildly restrictive. The size of the defect has been a more reliable measurement than the gradient across it to reveal an adequate but not too restrictive ASD. In some patients, depressed LV function will elevate the left atrial pressure and reduce transatrial gradient. Further tightening of the ASD snare in this setting may produce inadequate right-to-left shunting and a reduced cardiac output.

The snared ASD is adjusted off bypass to provide a small right-to-left gradient, which encourages TV flow. The atrial septum is shown with a right-to-left signal crossing the snare-adjusted atrial septal defect, which is about 3 mm in diameter (yellow arrows). Flow (white arrow) takes a mildly angulated course through the snared tissue of the fossa ovalis (A).

Flow is continuous when the atrial septal defect is restrictive. The gradient is assessed from the continuous wave spectral Doppler flow signal aligned with the color jet. (Here the continuous flow signal with a 1 m/s peak velocity and a 0.65 m/s mean velocity indicates a 1.7 mmHg mean pressure gradient) (B). LA = left atrium; RA = right atrium.

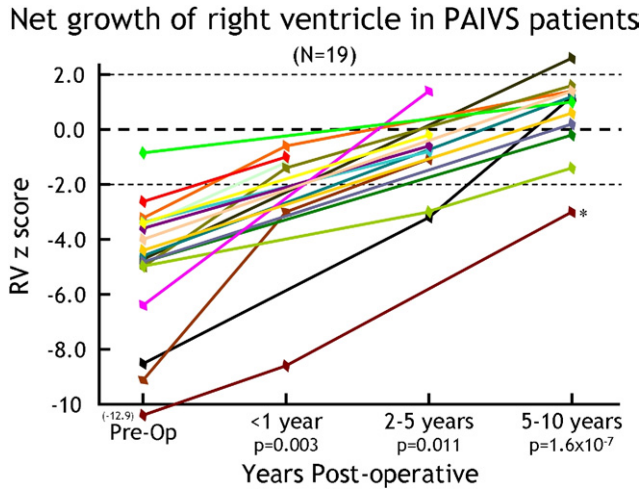


Figure 13 RV graph. The net growth of RVs in PAIVS patients is plotted (N = 19). To provide accurate individual data, paired biplane echo studies of RV volumes were compared following the growth procedure. The paired results of follow-up studies of net individual growth (calculated as z scores, the difference in standard errors of the mean from expected) of RVs are plotted as colored lines. Significant net growth was present at the earliest time point (<1 year) and continued to increase at 2 to 5 years and 5 to 10 years. (*Denotes patient with incomplete relief of RVOT obstruction and 2 large ASDs.) PAIVS = pulmonary atresia with intact ventricular septum; RV = right ventricle. (Reprinted with permission from Foker et al.[12])

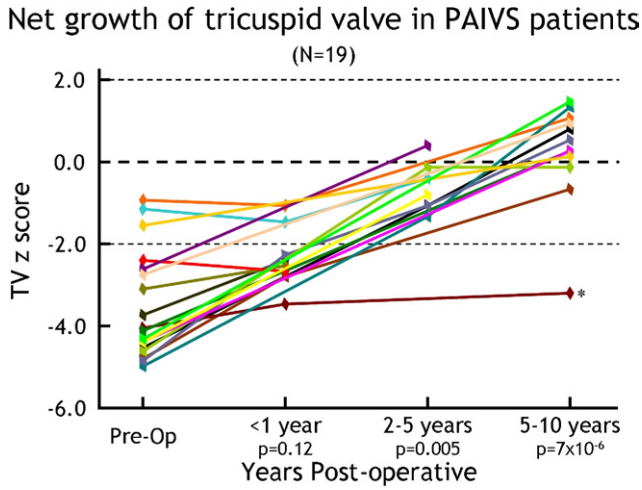


Figure 14 TV graph. Paired individual studies are plotted to show the net growth of TVs in PAIVS patients (N = 19). Again, net growth was assessed by z scores of the TVs and is shown by individual lines between the paired studies. Although trans-TV flow increased quickly as assessed by Doppler-echo (data not shown), the valve diameters lagged behind and did not show a significant increase in the less than 1-year follow-up studies. The normal diameter range was reached by 2 to 5 years. (*Denotes patient with incomplete relief of RVOT obstruction and 2 large ASDs; without an effective growth stimulus the TV did not reach normal TV size.) PAIVS = pulmonary atresia with intact ventricular septum; TV = tricuspid valve. (Reprinted with permission from Foker et al.[12])

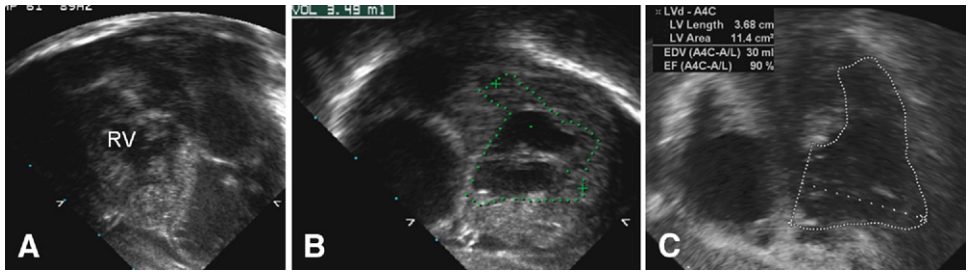


Figure 15 An example of RV growth after ligation of 6 RV-CACs, RV decompression, and snaring of the ASD.

(A) The preoperative apical 4-chamber view of the RV shows a very hypoplastic cavity surrounded by hypertrophied muscle. Both the TV and the mitral valves were also essentially covered by atrial tissue, which greatly compromised cardiac output. Both ventricles had an end-diastolic pressure of 24 mmHg at preoperative cardiac catheterization.

(B) The long-axis view similarly shows significant cavitary hypoplasia surrounded by hypertrophied RV myocardium with an obliterated infundibulum. By biplane analysis, the RV had a z score of -7.0. Off bypass, before repair, 6 RV-CACs were ligated.

(C) After 2 years the RV has a normal contour and, despite the initial poor condition of the myocardium, the volume has an essentially normal z score of -2.1 and an ejection fraction of 90%, presumably from the pulmonary regurgitation. The shunt had closed. RV = right ventricle. (Color version of figure is available online at http://www.optechtcs.com.)

tion abnormality (WMA), the tie can easily be removed. Finally, as discussed, ligation of the RV-CACs and relief of the RV outflow tract obstruction should be beneficial whatever repair endpoint is reached.

Conclusions

Our treatment plan, which has been applied to all PAIVS patients with proximal aortic-coronary artery continuity, has been to ligate the significant RV-CACs, completely relieve RVOT obstruction, and reduce the ASD by a snare to encourage TV flow. Although close cooperation by the team is required to achieve these results, the operation provides better myocardial perfusion, relieves the RV hypertension, and does not seem to be intrinsically destabilizing. Nevertheless, there are important postoperative considerations.

The antegrade flow after the connections are ligated improves the situation; however, the consequences of hypoxia are not quickly reversible. Significant diastolic dysfunction should be anticipated and, in some cases, even LV systolic function is abnormal. Postoperative extracorporeal membrane oxygenation (ECMO) support has been required in several cases.

Judging the degree of restriction for the snared ASD may pose difficulties. For severe cases, as noted, there may be preoperative instability with significant left ventricular dysfunction with elevated left atrial pressures from the preoperative retrograde hypoxic perfusion. The ASD may be reduced too much when based on the right-to-left pressure gradient and the right-to-left shunt will not be adequate to maintain a good cardiac output. We therefore do not recommend an ASD smaller than 3 to 4 mm to insure a satisfactory cardiac output and avoid excessively high right atrial pressures. As the patient stabilizes and begins to grow, the ASD will encourage more TV flow.

Not all of the RV-CACs may be apparent at the time of ligation. Those judged to be insignificant and not ligated did not enlarge or produce dysfunction. For 3 other patients, however, apical-septal dysfunction showed up 2 hours to 2 days later. The apical-septal segment is the last portion supplied by coronary blood flow and is where dysfunction occurs even though the steal site may be considerably upstream. Presumably, the connections were there preoperatively but without discernible flow. The significance of the late-appearing connections was quite variable and ranged from requiring ECMO support and ligation at reoperation to being an incidental follow-up finding.[9]

Consequently, after the initial ligation of our RV-CACs, we recommend follow-up echocardiogram about 2 hours after the procedure, on the first and second postoperative days, and any time ventricular function appears to worsen. Although this does appear to add another level of complexity to these patients, it requires no new diagnostic or operative challenges.

The reason ligating the RV-CACs had no apparent adverse consequences in our series may be because the myointimal lesions have developed over a number of months and adaptations by collaterals and other accommodations will have taken place. The vessels distal to the RV-CACs and with intimal lesions may appear falsely narrow because the connections may make the proximal coronary artery unusually large. The presence of the RV-CACs and the resulting retrograde coronary perfusion with hypoxic blood, however, has been shown to be detrimental.[4,7] Consequently, ligation of the RV-CACs appears beneficial no matter what final repair is achieved.

Currently, there are no other methods to eliminate the RV-CACs and allow a 2VR. The vessels and the infants are too small for catheter-based techniques and elimination of the RV cavity or closure of the TV, at best, insures a SVR.

Our series suggests that RV-CAC ligation can be readily done and results in antegrade flow with improved myocardial oxygenation. Presumably, the myointimal lesions may resolve and RV mechanics will continue to improve. A longer-term follow-up of our patients has shown RV catch-up growth in all and even the subset of very small TVs ($z \leq -4.0$) grew to normal size with the 1 exception being a patient lost to follow-up for several years who had 2 large ASDs and moderate RV outflow obstruction (Figs. 13-15). RV function in these patients, moreover, was in the normal or supranormal range because of the wide open pulmonary valve area.[9] One caveat is that about 20 to 25% of the TVs at the severe end of the spectrum have a structural abnormality, usually stenosis, in addition to the hypoplasia, although TV replacement has not been required in our series.[9]

These children resemble well repaired tetralogy of Fallot and the main remaining defect for these patients is the absence of a pulmonary valve. It is likely that a number of them will require pulmonary valve placement sometime in their young adult life. In summary, these results predict a substantial long-term advantage over those who undergo a SVR and, although questions remain, the significant long-term benefits of a 2VR seem achievable in almost all cases.[8,12]

Supplementary Data

Supplementary data associated with this article can be found, in the online version, at doi:10.1053/j.optechstcvs.2010.07.003.

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