Diagnosis and Management of Right Ventricle–Dependent Coronary Circulation in Pulmonary Atresia With Intact Ventricular Septum

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Background. Coronary artery anomalies including 1) right ventricle (RV)–to–coronary artery fistulas, 2) coronary artery stenoses, and 3) coronary occlusions occur in patients with pulmonary atresia with intact ventricular septum (PA-IVS). In some, a large part of the coronary blood supply may depend on the RV. This RV-dependent coronary circulation may determine survival after right ventricular decompression (RVD); RVD may cause RV “steal” in the presence of fistulas alone and ischemia, coronary isolation, or myocardial infarction in the presence of coronary stenoses.

Methods and Results. Eighty-two patients with PA-IVS who presented between January 1979 and January 1990 were reviewed; 26 (32%) had RV-to-coronary artery fistulas. Of these 26, 23 had adequate preoperative coronary angiograms for analysis. RVD was achieved in 16. Seven of 16 had fistulas only; each survived RVD. Six of 16 had stenosis of a single coronary artery [left anterior descending coronary artery (LAD), four; right coronary artery (RCA), two]; four of six survived RVD. Three of 16 had stenoses and/or occlusion of both the RCA and LAD; all three died shortly after RVD of acute left ventricular dysfunction.

Conclusions. 1) Potential RV steal alone does not preclude successful RVD. 2) Fistulas with stenoses to a single coronary artery may not preclude successful RVD. 3) RVD appears to be contraindicated in the presence of stenoses and/or occlusion involving both the right and left coronary systems. Nonsurvival after RVD seems to depend on the amount of the left ventricular myocardium at risk, i.e., that which is distal to coronary artery stenoses, especially when involvement of both coronary arteries limits effective collateralization. Precise definition of coronary arterial anatomy is mandatory in neonates with PA-IVS.

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Key Words • congenital heart disease • right ventricle • fistulas • surgery, heart

D ecisions regarding the surgical management of pulmonary atresia with intact ventricular septum (PA-IVS) have focused primarily on the size of the right ventricle (RV)1–20 and on the number of constituent right ventricular parts (inflow, trabecular, outflow).6,11,14,18,21–30 Although most workers have abandoned a biventricular repair in the presence of a diminutive RV,6,9,10,14,17,19,20,24,30–34 occasional reports14,24,35,36 have documented the capacity of even the smallest RVs to grow.

With the emphasis on cavity size and anatomy, the associated problems of RV-to–coronary artery fistulas and coronary stenoses have only recently received attention as determinants of surgical management.7,19,20,25,29,31,33,34,37–57 Because the suprasystemic RV may supply a substantial fraction of myocardial blood flow in the presence of fistulas with or without coronary stenoses, surgical procedures that result in RV decompression (RVD) may potentially compromise perfusion of various amounts of myocardium. In an effort to focus on this problem, the term “right ventricle–dependent coronary circulation” (RVDCC) has been introduced.31,33,34,54,57

The potential mechanisms for adverse outcome after RVD may depend on the specific coronary anatomy (Figure 1): 1) In patients with RV-to–coronary artery fistulas without coronary stenoses, RVD could result in a right ventricular “steal” phenomenon, i.e., runoff from the aorta into the RV during diastole.25,33,34,37,38,40,42–44,48,52,53

2) In patients with RV-to–coronary artery fistulas with coronary stenoses, RVD could lead to a right ventricular steal if the stenosis is distal to the fistulas and steal and/or ischemia if the stenosis is proximal to the fistulas.

3) Finally, in patients with RV-to–coronary artery fistulas with coronary occlusion or atresia proximal to a fistulous insertion, RVD could result in coronary ischaemia (i.e., RV-dependent supply of the myocardium distal to the occlusion) and myocardial infarction. In an at-
Figure 1. Diagram of potential mechanisms for adverse outcome with right ventricular decompression in patients with pulmonary atresia with intact ventricular septum and right ventricle-to-coronary artery fistulas. Ao, aorta; RCA, right coronary artery; RV, right ventricle.

Figure 2. Flow chart of surgical management in 26 patients with pulmonary atresia with intact ventricular septum (PA/IVS) and right ventricle-to-coronary artery fistulas. RVD, right ventricular decompression.

Figure 3. Diagrams of locations of fistulas in 23 patients with pulmonary atresia with intact ventricular septum. Top panel: Diagrammed on the left is the right coronary artery (RCA) in the left anterior oblique view and on the right, the left coronary artery (LCA) in the right anterior oblique view. Circled numbers represent the number of fistulas seen to the proximal, mid, posterior descending, and distal RCA; to the proximal, mid, and distal left anterior descending (LAD); and to the circumflex (Cx) coronary arteries, respectively. Bottom panel: Location of stenoses and occlusions in 23 patients. Stenoses are noted by the solid bars and occlusions by the dashed lines.
Methods

Eighty-two patients with the diagnosis of PA-IVS presented to The Children’s Hospital, Boston, between January 1979 and January 1990. All patients underwent cardiac catheterization with angiography on presentation. Review of these angiograms demonstrated 26 of the 82 (32%) to have RV–to–coronary artery fistulas. A fistula was defined as a patent communication between the RV and a coronary artery. Right ventriculography was available for review in all patients. In addition to right ventriculography, the coronary circulation was imaged by left ventriculography (16 of 23), aortography (12 of 23), balloon occlusion aortograms (seven of 23), and/or selective coronary injections (seven of 23). The coronary circulation was adequately imaged in 23 of 26.

Twenty of the 23 patients presented in Boston for initial management; three had initial shunt procedures elsewhere. RVD was attempted in 19 of the 23 (83%) and was deferred in four. In five of 19, RVD was inadequate, as documented by subsequent catheterization showing sustained suprasystemic right ventricular pressure. In two of these five, a second attempt at RVD was successful. Therefore, RVD was eventually adequate in 16 patients (Figure 2). Angiograms before RVD were reviewed, and the correlation between coronary anatomy and survival after RVD was assessed from these 16 patients.

In six patients, adequate serial angiograms of the coronary arteries in the presence of sustained suprasystemic right ventricular pressure were available for review (18 angiograms in six patients). This group included three patients in whom RVD was initially unsuccessful, one patient in whom RVD was deferred, and two patients who had initial shunt procedures elsewhere. The progression of coronary disease with sustained right ventricular hypertension was assessed from these six patients.

Figure 4. Angiograms showing fistulas with stenoses/occlusions of both the right and left coronary systems (patient 15). Top panel: Right ventricular (RV) injection in the right anterior oblique view (RAO) demonstrates fistulas (F) to the proximal and mid left anterior descending (LAD) coronary arteries with occlusion (arrow) between the fistulas (F1 and F2). The LAD continues beyond F2. Prominent RV apical vessels (AV) extend to the left ventricular apex and inferior wall. Bottom panel: Selective left coronary injection in the left anterior oblique view (LAO) opacifies the left circumflex coronary artery (Cx), which collateralizes to the right coronary artery (RCA), which is proximally occluded. The LAD is occluded beyond F1. This patient died after right ventricular decompression.
In five patients, serial angiograms subsequent to RVD were available for review (12 angiograms in five patients).

Results

Angiographic Anatomy

The locations of fistulas and stenoses in the 23 patients reviewed are shown in Figure 3 and Table 1. Seven had fistulas only (30%), and 16 had stenoses in addition to fistulas (70%). Fistulas were most commonly connected to the proximal or mid left anterior descending (LAD) and the mid right coronary artery (RCA). Eighteen patients had more than one fistula, and 10 patients had more than one fistula to a given coronary artery. A fistula to the circumflex artery (Cx) was seen in only one patient (Figure 3).

Stenoses occurred most commonly in the mid LAD. Eight patients had more than one stenosis, and four patients had more than one stenosis in a given coronary artery. Stenoses were most commonly found in close proximity to fistulous insertions, being either immediately proximal or distal to the fistula. A stenosis not in close proximity to a fistulous insertion was seen in only one patient (Figure 3).

Five patients had a complete coronary artery occlusion demonstrated at initial catheterization. In three, the proximal RCA could not be visualized, and differentiation between occlusion and absence of the proximal segment was not possible. Two of these three had an additional occlusion of the mid or distal LAD. Other coronary arterial abnormalities in addition to fistulas and stenoses included dilated coronary arterial segments (11 patients), coronary artery collaterals (six patients), and single coronary artery (one patient).

An unusual pattern of right ventricular vascular supply to the left ventricle was seen in seven patients. The right ventricular apex gave off several "vessels" in a spraylike arrangement to the left ventricular apex, inferior wall, and/or posterior wall. These right ventricular apical vessels did not have the form or distribution of normal coronary arteries or collaterals and appeared to be supplying areas of the left ventricular myocardium that had decreased coronary perfusion secondary to a coronary artery stenosis or occlusion of one of the native coronary arteries (Figures 4–7).

RV Morphology

All patients except three (patients 14, 22, and 23) were thought to have a tripartite RV by angiography (inflow, trabecular portion, outflow). Some degree of tricuspid regurgitation was present in all patients in whom it could be assessed (Table 1). RV size was estimated to be severely hypoplastic in 14, moderately hypoplastic in eight, and normal in one. Tricuspid valve diameters calculated from right ventriculograms were listed (Table 1).

Relation Between Coronary Artery Anatomy and Outcome After RVD

Of the 23 patients who are the subject of this angiographic review, 16 underwent RVD sufficient to decrease the right ventricular pressure below one half the systemic pressure. The median age at RVD was 5 days (range: 2 days to 33½ years; mean, 6.7 months). Of this group of 16 patients, 11 survived and five died.

The surgical procedures used were varied and included transannular pericardial outflow tract patch plus Blalock-Taussig shunt (seven patients), outflow tract patch without shunt (two patients), outflow tract patch after an initial shunt procedure (two patients), RV–to–pulmonary artery homograft conduit after an initial shunt procedure elsewhere (two patients), outflow tract patch after initial pulmonary valvotomy plus shunt (two patients), and tricuspid valvectomy plus formalization of the ductus arteriosus (one patient) (Table 1).

The 16 decompressed patients were divided into three groups based on coronary anatomy (Figure 8): group 1, fistulas without coronary stenoses; group 2, fistulas with stenoses in either the right or left coronary arteries; and group 3, fistulas with stenoses involving both the right and left coronary arteries. There were seven patients in the first group (fistulas without coronary stenoses). All seven survived RVD (Figure 8, Table 1). Fistulas connected to both the RCA and the LAD in three patients, to the RCA alone in two patients, and to the LAD alone in one patient. The seventh patient, who had anomalous RCA origin from the LAD, had a large fistula to the mid LAD (Figure 9). Two of the seven patients in this group had fistula ligation at the time of RVD. Coronary collaterals were not seen in any of these patients without coronary stenoses.

There were six patients in the second group (fistulas with stenosis of either the right or left coronary artery, but not both). All six patients survived the surgical procedure. Two patients died, however, in the postoperative period, one with and the other without evi-
Table 1. Patient Characteristics

Characteristics of 16 patients undergoing RVD

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<th>Cx</th>
<th>Collaterals</th>
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RVD, right ventricular decompression; RCA, right coronary artery; LAD, left anterior descending; Cx, circumflex; RV, right ventricle; TV, tricuspid valve; TR, tricuspid regurgitation; F/U, follow-up; m, mid; F, fistulas; p, proximal; nl, normal; sm, severely hypoplastic; RVOT, RV outflow tract patch; RMBTs, right modified Blalock-Taussig shunt; ASD, atrial septal defect; A, alive; d, distal; md, moderately hypoplastic; LMBTS, left modified Blalock-Taussig shunt; PA, pulmonary artery; S, stenosis; D(OR), ICU death; UE, unable to evaluate; Pul, pulmonary; RPA, right pulmonary artery; O, occlusion; formalin PDA, formalin infiltration of patent ductus arteriosus; D(OR), death in operating room; VSD, ventricular septal defect; LPA, left pulmonary artery; AO, aorta; D, dead.

*Incomplete RVD (RV pressure > 1/2 systemic pressure).

dence of left ventricular dysfunction (Figure 8, Table 1). Patient 8, who had a single stenosis of the mid LAD, died on day 2 after surgery of respiratory complications without clinical or echocardiographic evidence of left ventricular dysfunction. Autopsy revealed a mild stenosis of the mid LAD without evidence of myocardial infarction. The second death in this group occurred on day 4 after surgery in an infant with a distal RCA stenosis (patient 13). This infant had signs of left ventricular dysfunction in the early postoperative period. His course was complicated, however, by prematurity, an intraventricular hemorrhage, and Blalock-Taussig shunt revision with subsequent thrombosis, making it difficult to pinpoint the exact cause of death. The coronary artery anatomy of the four survivors of this group included stenosis of the distal RCA in one patient and stenosis of the proximal and/or mid LAD in three. Two of the survivors each had two discrete stenoses of the LAD (patients 11 and 12).
Of the six decompressed patients with stenoses of a single coronary artery, collateral vessels were seen in two, both survivors. In patient 10, the LCx collateralized to the LAD, which was stenotic in its midportion. In patient 11, the RCA collateralized the LAD in the presence of proximal and mid LAD stenoses (Figures 10 and 11). Three of the patients in this group (patients 8, 11, and 13) had stenosis proximal to a fistulous insertion, a situation where there is potential for coronary isolation, i.e., RV-dependent supply of the entire myocardial territory distal to the stenosis. Of these three, only the patient with coronary collaterals (patient 11) survived.

There were three patients in the third group (fistulas with stenoses and/or occlusion involving both the RCA and LAD). All three died shortly after RVD, two in the operating room and the third 18 hours after surgery. Two of the three (patients 14 and 15) had not only stenosis but also occlusion of both the RCA and LAD, resulting in, first, potential isolation of both coronary systems and, second, severe reduction in the potential for adequate collateralization of one system to another. One of the three (patient 14) had a stenosis of the proximal LCx as well (Figure 8, Table 1). Coronary artery collaterals were present in two of the three in this group and included a LCx collateral to a proximally occluded RCA (patient 15) and a LAD collateral into LCx territory where the proximal LCx could not be seen (patient 16). Interestingly, the unusual, spraylike vessels emanating from the RV apex described above were
FIGURE 6. Angiograms showing fistulas with stenoses/occlusions of both the right and left coronary systems (patient 22). Top panel: Right ventricular (RV) injection demonstrates two fistulas (F) to the mid right coronary artery (RCA) with occlusion (arrow) between the fistulas. A fistula to the proximal left anterior descending coronary artery (LAD) is also opacified. LAO, left anterior oblique. Bottom panel: The right anterior oblique (RAO) view demonstrates the fistulas to the RCA and LAD. The proximal LAD is occluded. The distal LAD is fed only by RV apical vessels (AV). Right ventricular decompression was deferred in this patient.

present in all three patients and may have been the sole supply to the apical myocardium in jeopardy because of more proximal stenoses (Figures 4–7).

Coronary Arteries in the Presence of Sustained Suprasystemic Right Ventricular Pressure

Six patients with sustained suprasystemic right ventricular pressure had serial angiograms adequate for review (patients 4, 9, 10, 15, 17, and 22). The mean duration of documented sustained suprasystemic right ventricular pressure was 36 months (range, 11 months to 10 years; median, 18.5 months). In four patients, there was no change in either the size or number of fistulas or in the existing coronary stenoses. In patient 4, who had a large fistula to the mid LAD and marked ectasia of the left coronary, serial angiograms over a 3-year period revealed increased ectasia of the proximal LAD and increased tortuosity of the fistula. In patient 22, who had a mid LAD stenosis demonstrated at 1 year of age, subsequent angiograms at 2.5 years of age failed to opacify the distal LAD, suggesting progression of the stenosis to an occlusion. In addition, right ventricular apical vessels extending to the left ventricular apex, although present on the initial study, became more prominent during the 1.5-year interval (Figures 6 and 7).

Fate of Fistulas and Stenoses After RVD

The mean angiographic follow-up after RVD in five patients (patients 1, 3, 9, 10, and 11) was 30 months (range, 6–49 months). In all five, right ventriculograms no longer opacified the coronary arteries. Fistulas remained completely open in patients 9 and 10, as shown by retrograde flow into the RV on aortic and/or selective coronary artery injection at 49 and 31 months after
FIGURE 7. Line drawing of Figure 6. Coronary occlusions shown by dashed lines. LAO, left anterior oblique; RAO, right anterior oblique; LCA, left coronary artery; F, fistula; RCA, right coronary artery; RV, right ventricle; LAD, left anterior descending coronary artery; AV, apical vessels.

RVD, respectively. In patient 1, selective coronary angiography barely opacified only one of what were multiple fistulas; the RV was not opacified. In patient 11, right ventriculography opacified the proximal portion of what was a fistula to the distal LAD without opacifying the LAD. In patient 3, a previously documented small fistula to the distal RCA opacified neither on right ventriculogram nor aortogram 6 months after RVD.

Three of five decompressed patients with serial angiograms had coronary artery stenoses demonstrated before RVD. In none of these five did the follow-up angiograms show new stenoses. Selective left coronary artery injection in patient 11 at 28 months after RVD, however, revealed progression of a mid LAD stenosis to an occlusion. Also demonstrated at this follow-up catheterization were new collaterals from the RCA to the distal LAD territory, with the maintenance of normal left ventricular function angiographically.

Long-term Follow-up

Among the 11 patients who survived RVD, there were no late deaths, with a mean follow-up of 22 months (range, 1–70 months). Four have had complete two-ventricle repairs; in three, the atrial septal defect and shunt are still open, and in two, the atrial septal defect is open with shunt closed. In the remaining patient (patient 10), the distal end of the right pulmonary artery was connected to the superior vena cava, with antegrade RV flow supplying the left pulmonary artery. Thus, in 10 of 11 patients, the RV provides most or all of the pulmonary blood flow. All 11 patients are alive and well.

Discussion

Relation of Survival After RVD to Coronary Anatomy

RV-to-coronary artery fistulas with and without coronary artery stenosis have been described in both necropsy and angiographic reviews of PA-IVS.19,20,25,29,31,33,34,36–40,44–63 It has been postulated from

FIGURE 8. Bar graph showing survival after right ventricular decompression (RVD). RCA, right coronary artery; LAD, left anterior descending coronary artery; CX, left circumflex coronary artery; ICU, intensive care unit; LV, left ventricular.
these studies that in the presence of a right ventricular steal phenomenon or in the presence of any or selective coronary artery stenoses, RVD would prove fatal. Despite this hypothesis, the relation between preoperative coronary artery anatomy and survival after RVD has remained controversial. Our bias toward decompressing every RV, regardless of size, has allowed us to assess the relation between coronary anatomy and the outcome of RVD. In our series of 16 decompressed patients, all seven patients with fistulas and no coronary stenoses survived. Six others with fistulas and stenoses of a single coronary artery initially survived RVD, but two of six died during the first week after surgery, one with and the other without evidence of left ventricular dysfunction. All three patients with fistulas and stenoses and/or occlusions involving both the RCA and LAD died shortly after RVD.

These observations seem to support several inferences. First, although fistulas without associated coronary artery stenoses may theoretically lead to a coronary steal into these small, noncompliant right ventricles, their presence does not preclude successful RVD. Several authors have cautioned the potential problem of right ventricular steal after right ventricular decompression, especially in the presence of proximal/mid as opposed to distal/apical fistulas. This concern was not substantiated in our series.

Second, fistulas with stenoses of a single coronary artery do not preclude successful RVD. Patients with stenoses of a single coronary artery, however, had a less favorable outcome. Four of six patients are currently alive and well after RVD. Of the two patients who died, only one had evidence of left ventricular dysfunction, and both had clinical circumstances that may have led to death irrespective of the coronary anatomy. It is noteworthy that three of the four survivors of this group had one or more proximal and/or mid LAD stenoses. Although several authors have cautioned against RVD in
FIGURE 10. Angiograms showing fistulas with stenosis to a single coronary artery (patient 11). Top panel: Aortic injection demonstrates an ectatic right coronary artery (RCA). Bottom panel: Right ventricular (RV) injection opacifies a fistula (F) to the mid left anterior descending coronary artery (LAD) with stenosis (arrow) just proximal to the fistulous insertion. The LAD is irregular proximally with an additional proximal stenosis (arrow). The RCA fills from collaterals to the LAD. This patient survived right ventricular decompression.

the presence of a proximal or mid LAD stenosis, we postulate that decompression may be tolerated as long as the RCA and Cx are uninterrupted so that there is adequate potential for collateralization. Coronary collateralization to a stenosed LAD was documented in two of the three survivors with LAD lesions.

Third and most important, RVD may be contraindicated in the presence of stenosis and/or occlusion involving both the right and left coronary systems. The three patients in our series who died immediately after RVD had stenoses and/or occlusion of both the RCA and the LAD. In addition to having involvement of multiple vessels, the patients in this group were the only ones to have complete coronary artery occlusions and/or proximal vessel atresia in addition to stenoses. Two of three had isolation of both coronary systems (almost entire coronary supply RV dependent), and the potential for isolation of the LAD existed in the third.

In retrospect, each of these patients had a major portion of the left ventricular myocardium dependent on supply from the RV and therefore at risk for ischemia when the RV was decompressed. In the presence of obstruction of both the right and left coronaries, the potential for effective collateral formation may be severely limited. Death after RVD seems to depend on the amount of left ventricular myocardium at risk for ischemia, namely that which is distal to coronary artery stenoses in the presence of stenoses in other vessels that prevent effective collateralization.

Late Coronary Changes With and Without RV Decompression

Although patient numbers are limited, our series gives evidence that in the face of sustained suprasystemic RV pressure, existing coronary stenoses and dilation may progress. Likewise, there is evidence that
existing coronary stenoses may progress even after the RV has been adequately decompressed. We did not, however, demonstrate the development of new stenoses either in the presence of sustained suprasystemic RV pressure or after successful RVD. Also, in contrast to published reports stating that after successful RVD, fistulas seem to involute and close in most, folllow-up angiograms after RVD in our series showed fistulas to be completely or partly patent in four of five patients.

**RV Size and Coronary Anatomy**

Severely hypoplastic and moderately hypoplastic RVs were present in all three coronary groups and in both survivors and nonsurvivors. In contrast to previously published work, in our series a tripartite RV was seen in the vast majority of patients with PA-IVS, although in several patients, the trabecular and outflow portions were quite small.

Although this study clearly underlines the importance of coronary anatomy in determining surgical management of patients with PA-IVS, several limitations must be noted. This is a rare disease, and the number of patients seen was relatively small. In addition, adequate coronary definition became routinely available only after 1984, and therefore 15% of the patients were excluded from one or more analyses. Finally, despite an institutional bias to decompress every RV regardless of size, decompression was not successful in every case. The loss of these patients from the analysis does not seem to alter the inferences, but this effect cannot be excluded.

**Conclusions**

Despite these limitations, several conclusions seem warranted: 1) Potential right ventricular steal, i.e., fistulas without associated coronary artery stenoses, does not preclude successful RVD. Procedures designed to minimize the effects of aortoventricular steal (i.e., thromboexclusion of RV) would thus seem superfluous or even dangerous. 2) Fistulas associated with stenoses to a single coronary artery may not preclude successful RVD, although this is a difficult group that warrants further investigation. 3) RVD appears to be contraindicated in the presence of stenosis or occlusion/occlusion/occlusion involving both the right and left coronary systems. Death after RVD seems to depend on the amount of left ventricular myocardium at risk for ischemia if the RV is decompressed, namely, that which is distal to coronary artery stenoses/occlusions, especially when involvement of both coronary arteries interferes with effective collateralization. 4) In the presence of RV-coronary artery fistulas, adequate definition of the coronary anatomy in the neonatal period (by angiography of the RV in addition to antegrade opacification of the coronary arteries) is necessary before RVD. 5) Because progression of coronary artery stenosis to occlusion may occur either in the presence of sustained suprasystemic right ventricular pressure or after RVD, close follow-up of stenotic lesions is needed.

In general, these data provide support for the concept that decisions regarding RVD in PA-IVS should be based, to a large extent, on coronary anatomy. They do not indicate, however, that RV outflow tract reconstruction is in fact warranted in all infants without a RVDCC. Such a conclusion could only be based on studies demonstrating RV growth regardless of RV anatomy. Clearly, continued follow-up in a larger group of patients is necessary.

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