Dilation angioplasty of congenital or operative narrowings of venous channels

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ABSTRACT Balloon dilation angioplasty was attempted in 10 infants and children with severe congenital or operative "venous" obstructions. In five children the obstructions were "vena caval" and followed repair for transposition of the great vessels (four patients) or orthotopic liver transplantation (one patient). In the four patients with fixed vena caval or baffle obstructions, balloon angioplasty was successful in relieving the obstruction, decreasing the average gradient (16.0 to 4.5 mm Hg), and increasing the average diameter (3.0 to 8.9 mm) of the obstructed site. One child died 1 week later from an unrelated cerebral hemorrhage; the three survivors have had persistent clinical and angiographic improvement. The fifth child had severe systolic narrowing of the superior baffle limb caused by marked tricuspid regurgitation, which ballooned the superior limb of the baffle against the atrial roof. Angioplasty was unsuccessful in relieving this type of obstruction, which resolved with tricuspid valve replacement. Of the five infants with obstructed pulmonary veins, three had congenitally narrowed vessels associated with total anomalous pulmonary venous connection, one had acquired stenosis, and one had postoperative obstruction after repair of a mixed type of total anomalous pulmonary venous connection. Dilation was unsuccessful in all five patients, but for different reasons: in the congenitally narrowed veins, the waist in the balloon could not be eliminated, even with high dilating pressures; in the infant with acquired stenosis, the vein stretched but did not tear at low dilating pressures; and in the postoperative obstruction, angioplasty increased the diameter of the obstruction but did not increase flow to the affected lung. Venous obstructions in children are heterogeneous lesions, and the success of angioplasty in relieving such obstructions will depend not only on the technique used but also on the nature of the obstruction.


Ballooning dilation angioplasty has recently been found to be effective in certain forms of arterial and valvular narrowings in children.1–4 As the experience with this technique has increased, however, it has become clear that the type of lesion, the age of the patient, and the specific dilation technique all may be important variables in determining the success or failure of the procedure.

Dilation angioplasty has also been used in venous obstructions in children, but the results have been disappointing. Driscoll et al.5 found that dilation of three pulmonary veins was unsuccessful, while Waldman et al.6 reported failure of dilation in a "mid-cavity baffle" obstruction in a child who had previously undergone Mustard's repair of transposition of the great vessels. Rocchini et al.,7 who reported a possibly successful superior vena caval dilation, despite use of a small (6 mm) balloon, were unable to provide meaningful follow-up data because of the death of the child 14 hr after angioplasty.

The reasons for the apparent failure of balloon dilation angioplasty in "venous" obstructions in these previous reports are not clear. Part of the uncertainty rests in the imprecision of the terms used to describe postoperative venous obstructions in children; thus, narrowings in or around the superior limb of a Mustard baffle have been variously termed "mid-cavity,"6 "superior vena caval,"8,9 and other similarly imprecise terms. In fact, the pathologic anatomy of these patients has not yet been carefully defined. Thus the term "caval" obstruction will be used in this article recognizing its limitations.

We have previously introduced the notion that with
compliant structures such as branch pulmonary arteries, very large balloons might be required for success. The balloon-induced enlargement in a vessel that occurs at low (1 to 2 atmospheres) dilating pressures may represent medial stretching (a result that provides ineffective long-term relief), whereas the enlargement occurring at higher pressures (≥5 atmospheres) is more likely to result in a medial tear, an event that appears to be necessary for a satisfactory long-term result.10, 11 With such an approach, we have attempted to use dilation angioplasty in the therapy of 10 consecutive infants and children with severe venous obstructions.

**Materials and methods**

Any patient who had signs of a major venous obstruction was considered a candidate for balloon dilation angioplasty. Those patients in whom surgical management was considered to be safe and effective (i.e., total anomalous pulmonary venous connection as an isolated lesion) were sent directly to operation. Those patients thought to be operable only at high risk, or to be inoperable, were offered an attempt at dilation angioplasty. Informed consent was obtained from the parents in every case. The protocol was approved by the Human Volunteers Committee of the University of Minnesota.

From October 1980 to September 1983, a total of 10 infants and children underwent attempted angioplasty. Nine of the 10 patients underwent dilation at one institution (University of Minnesota). The clinical characteristics, diagnoses, and vessels that were dilated are summarized in Table 1. The patients ranged in age from 2 days to 11 years and weighed from 3 to 56 kg. Five children had pulmonary venous obstructions, and five had caval obstructions. In eight of the 10 patients angioplasty was attempted during cardiac catheterization; venous entry was obtained percutaneously from either the femoral or internal jugular vein. General anesthesia (ketamine) was used when the internal jugular approach was used. In the child with inferior vena caval obstruction after liver transplantation, the procedure was performed percutaneously in the operating room with portable fluoroscopy because of concerns that vessels in patients in the immunosuppressed state might be particularly susceptible to rupture. Angioplasty of the left pulmonary veins (patient 4) was performed from a left thoracotomy after surgical isolation of the veins; prior surgical closure of the atrial septum in that child as an infant precluded the use of a transseptal approach.

Since no one had previously identified a successful protocol for the dilation of venous structures, we used an approach similar to that previously described for branch pulmonary arteries. A balloon whose inflated diameter was at least twice as large as the obstructed site was positioned appropriately and inflated to 1 to 2 atmospheres. We know from previous experience that such low pressures may stretch a vessel but are unlikely to produce a medial and intimal tear. If no waist was present at low inflating pressures, the balloon was removed, and progressively larger balloon diameters were used until a clear-cut waist was seen in the balloon at low inflating pressures. The pressure in the balloons were then increased for 10 to 30 sec until the waist disappeared, the balloon ruptured, or the pressure reached 5 to 8 atmospheres (see table 2). We never advanced an unguided catheter or guidewire across a previously dilated site; since successful dilations of other lesions produce intimal and medial tears, an unguided catheter may enter such a partial tear and produce vascular rupture.10 After angioplasty, repeat determinations of the diameter at the obstructed site, and the gradient across the site, were obtained when possible to determine the success or failure of the procedure. A procedure was considered successful if the diameter of the obstruction doubled and the gradient fell by more than 50%.

**TABLE 1**

Clinical summary

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Weight (kg)</th>
<th>Diagnosis</th>
<th>Previous operations</th>
<th>Vein dilated</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1 day</td>
<td>2</td>
<td>Single atrium, single ventricle, TAPVC to R SVC</td>
<td>None</td>
<td>Common pulmonary vein</td>
</tr>
<tr>
<td>2</td>
<td>2 day</td>
<td>3</td>
<td>Cor triatriatum, subaortic stenosis, TAPVC to LSVC</td>
<td>None</td>
<td>Common pulmonary vein</td>
</tr>
<tr>
<td>3</td>
<td>5 mo</td>
<td>5</td>
<td>Stenosis of the individual pulmonary veins</td>
<td>None</td>
<td>Right lower pulmonary vein</td>
</tr>
<tr>
<td>4</td>
<td>1 yr</td>
<td>5</td>
<td>TAPVC of mixed type: left veins to LSVC, right veins to coronary sinus</td>
<td>Complete repair</td>
<td>Left lower pulmonary vein</td>
</tr>
<tr>
<td>5</td>
<td>2 yr</td>
<td>10</td>
<td>Mitral atresia, double-outlet right ventricle, TAPVC to LSVC</td>
<td>Pulmonary arterial banding</td>
<td>Common right pulmonary vein</td>
</tr>
<tr>
<td>6</td>
<td>7 mo</td>
<td>7</td>
<td>Biliary atresia</td>
<td>Orthotopic liver transplant</td>
<td>Inferior vena cava</td>
</tr>
<tr>
<td>7</td>
<td>3 yr</td>
<td>11</td>
<td>Transposition of the great vessels</td>
<td>Blalock-Hanlon, Mustard</td>
<td>Superior limb of baffle</td>
</tr>
<tr>
<td>8</td>
<td>6 yr</td>
<td>17</td>
<td>Transposition of the great vessels, VSD</td>
<td>Mustard, VSD closure</td>
<td>Superior limb of baffle</td>
</tr>
<tr>
<td>9</td>
<td>11 yr</td>
<td>39</td>
<td>Transposition of the great vessels</td>
<td>Mustard, Dacron baffle, VSD closure</td>
<td>Inferior limb of baffle</td>
</tr>
<tr>
<td>10</td>
<td>12 yr</td>
<td>56</td>
<td>Transposition of the great vessels</td>
<td>Mustard, Dacron baffle</td>
<td>Inferior limb of baffle</td>
</tr>
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**TAPVC** = total anomalous pulmonary venous connection; **RSVC** = right superior vena cava; **LSVC** = left superior vena cava; **VSD** = ventricular septal defect.
TABLE 2

<table>
<thead>
<tr>
<th>Techniques of balloon angioplasty</th>
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<tr>
<td></td>
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<tr>
<td><strong>Patient</strong></td>
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sys. = systole; dias. = diastole.

^Balloon ruptured.

After dilation, all children were observed in the intensive care unit overnight. Those children with apparently successful procedures were discharged the following day, and low-dose aspirin therapy was added to their regimen (5 to 10 mg/kg/day) for 1 to 3 months. Predilation and postdilation two-dimensional echocardiograms were obtained for every patient to provide a basis for noninvasive follow-up treatment. However, precise sizing of the veins by echocardiographic techniques proved difficult. In the two children with inferior vena caval obstruction, the size of the hepatic veins was easily determined before dilation and was used to estimate the degree of elevation of inferior vena caval pressure before and after dilation. Repeat catheterization was performed in three patients 4, 6, and 7 months after dilation.

Results

Pulmonary veins. Of the five children with obstructed pulmonary veins, three had congenital narrowings of anomalous pulmonary veins (see table 1). In two of these infants, angioplasty was attempted in the first 2 days of life to try to reverse unremitting hypoxemia, acidosis, and shock. In all three cases of congenital pulmonary venous stenosis, angioplasty was unsuccessful for the same reason; despite use of the appropriate balloon, correctly positioned, the waist could not be eliminated even in the face of very high dilating pressures (figure 1). In one infant (No. 1) the inflation of an older polyvinylchloride balloon to high pressure caused the balloon to distend above its nominal diameter, causing a venous tear and a modest mediastinal hemorrhage. All three children with congenital venous narrowings died 1 to 90 days after attempted angioplasty. In each, death appeared to be caused by the unrelieved pulmonary venous obstruction and was unrelated to the attempted angioplasty. Examination of the postmortem specimens at the site of obstruction revealed prominent intimal proliferation. The intima was composed of collagen and disorganized elastic fibers. The boundary between media and adventitia was usually indistinct. The vein wall thickening at the site of obstruction was composed largely of collagen. Elastic fibers tended to be sparse and diffusely scattered throughout the vessel wall. In these infants there was no evidence for venous wall tears at the site of obstruction.

One infant, who had obstruction of the individual pulmonary veins, was asymptomatic until she presented with respiratory distress at 4 months of age. Because of the advent of symptoms so late in infancy, she was presumed to have had "acquired" narrowings. The right upper vein was found to be occluded at initial catheterization; the right lower vein was severely stenotic, and the left veins were mildly narrowed angiographically. Dilation of the right lower vein with a balloon 3½ times the narrowed site did not produce a visable waist even at low pressures. Although there was a transient fall in main pulmonary arterial pressures from 15 mm Hg above systemic pressure levels to systemic levels, dilation produced acute hemoptysis, necessitating emergency intubation. She recovered uneventfully from this procedure. Signs of severe pulmonary congestion did not resolve, and she died suddenly 1 month later while awaiting a second attempt at
An angioplasty catheter (table 2) was advanced over the guidewire, its position was confirmed with portable fluoroscopy, and the narrowing was dilated. The distal pulmonary vein was repaired at the needle entry site with 7-0 prolene sutures, and the chest was closed. Repeat cardiac catheterization 1 week later demonstrated considerable increase in the caliber of the narrowed site (figure 2), but two repeat lung scans 1 and 4 months later showed no improvement in the flow (9% and 7%) directed to the affected lung.

**Venae cavae.** Of the five children with vena caval or intracardiac baffle obstructions after the Mustard procedure, four had fixed obstruction in that the diameter of the narrowing did not change with the cardiac cycle.

dilation with a larger balloon. Postmortem examination revealed no evidence of an intimal tear in the right lower pulmonary vein. Arterioles and venules in both lungs demonstrated prominent medial hypertrophy.

The fifth infant had undergone successful repair of mixed-type total anomalous pulmonary venous connection at 2 weeks of age. However, growth failure was noted by 6 months of age, and serial cardiac catheterizations documented progressive obstruction of the left pulmonary veins at their entry to the left atrium. By 1 year of age the diameters of the left upper and lower pulmonary veins were 1 mm (figure 2), and only 9% of the total cardiac output was directed to the left lung as estimated by quantitative perfusion lung scan. At left thoracotomy, the left lower pulmonary vein was isolated surgically and punctured with a 19-gauge needle at its widest position; a 0.038 guidewire was advanced into the left atrium through the stenosed portion of the

![FIGURE 1](image1.png) **FIGURE 1.** Pre and postdilation angiograms of the common right pulmonary vein from patient 5. A 10 mm Medi-Tech angioplasty balloon has been inflated to 8 atmospheres, but the waist in the balloon did not disappear (**middle panel**).

![FIGURE 2](image2.png) **FIGURE 2.** A 7F end-hole balloon-tipped catheter has been positioned in the proximal left pulmonary artery. The balloon is inflated to occlude flow, and 7 to 10 ml of contrast is injected distal to the balloon and flushed through with saline (patient 4). The left lower pulmonary vein is opacified before and after dilation, with evidence of an improved diameter at the obstructed site.
In each of these children results of angioplasty appeared to be quite successful (figure 3), increasing the diameter of the narrowing by nearly threefold and decreasing the gradient by at least 60% (table 2). Very large balloons were required (figure 3) in each case, indicating that caval or intracardiac baffle obstructions are generally very compliant structures, stretching but not tearing over a large range of balloon diameters. In each case, however, a waist was eventually seen that disappeared at balloon pressures of 5 atmospheres. Regression of the signs of obstruction has persisted for 6 to 10 months in the three late survivors (table 2). Late echocardiographic results have demonstrated a 30% to 40% decrease in hepatic vein diameter in patient 9, in whom repeat cardiac catheterization has also been performed. Two patients demonstrated persistence of the obstruction relief (figure 4, table 3) at cardiac catheterization.

In the fifth child a 12 mm Hg gradient across the superior limb of the Mustard baffle was found to be dynamic; during systole the diameter of the baffle decreased to 2 mm, whereas during diastole it appeared to be unobstructed (figure 5). An attempt at dilation with a large balloon revealed only a subtle waist, which disappeared upon inflation. However, dilation resulted in little hemodynamic or angiographic improvement. Since the systolic baffle "obstruction" was associated with marked tricuspid (systemic atrioventricular valve) insufficiency and a 36 mm Hg "V" wave in the pulmonary venous portion of the atria, the child underwent systemic atrioventricular valve replacement, without correction of the baffle. This operation corrected his symptoms of "superior vena caval" obstruction. At operation, the superior baffle appeared to be unobstructed.

**Discussion**

Although balloon dilation of vascular narrowings in children is still a novel technique whose ultimate effectiveness remains to be documented, certain principles have emerged over the past 4 years of clinical use. The success or failure of the technique will depend on the type of lesion being dilated, the age of the patient, the size of the balloon relative to the diameter of the obstruction, and the pressure applied. Other variables that may or may not influence the success of angioplasty include the number and duration of dilations and/or the use of anticoagulants and anti-inflammatory agents. Not only is a careful catalog of angioplasty failures necessary to avoid their repetition but also documentation of the technique used during failures allows other workers to plan subsequent approaches to the problem. The purpose of this article was twofold:

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**TABLE 3**

Late results of dilation angioplasty of postoperative caval obstructions

<table>
<thead>
<tr>
<th>Patient</th>
<th>Immediate postdilation</th>
<th></th>
<th>Postdilation (6–7 mo)</th>
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<tbody>
<tr>
<td></td>
<td>Diameter (mm)</td>
<td>Gradient (mm Hg)</td>
<td>Diameter (mm)</td>
</tr>
<tr>
<td>9</td>
<td>14</td>
<td>0</td>
<td>12</td>
</tr>
<tr>
<td>10</td>
<td>12</td>
<td>4</td>
<td>11</td>
</tr>
</tbody>
</table>

**FIGURE 3.** Predilation and postdilation angiograms of the inferior vena cava in patient 9. Two angioplasty balloons were used to dilate the obstructed vessel.
FIGURE 4. Late (6 months after dilation) angiogram in patient 9.

FIGURE 5. Dynamic superior vena caval obstruction as seen before dilation during both systole and diastole in patient 8.
to report the success of angioplasty in relieving vena caval obstructions and its failure in relieving pulmonary venous obstructions, and to analyze the potential reasons for those successes and failures.

**Pulmonary veins.** Dilation of congenitally stenosed pulmonary veins in infants was unsuccessful in all three cases, and the reason for the failure is clear: the lesion was too rigid for the balloon to be able to eliminate the waist. The structural nature of a stenosed pulmonary vein that results in this rigidity is not known; obviously more extensive correlation of the results of angioplasty with the gross and microscopic nature of such lesions will be needed to identify why such lesions are undilatable. In any case, it is unlikely that progress will be made in dilating such lesions until balloons capable of withstanding higher dilating pressures become available.

The failure of angioplasty to produce a beneficial hemodynamic result in the child with stenosis of the left pulmonary veins after correction of total anomalous pulmonary venous connection raises a more difficult problem. Despite apparent anatomic improvement (figure 2), there was no increase in flow to the affected lung. This finding suggests that a prolonged decrease in flow to one lung, produced by unilateral venous obstruction, can result in structural abnormalities in the vascular tree of the obstructed lung that may not be reversible. If such is the case, angioplasty will be required in the early postoperative period to “protect” the pulmonary vascular tree.

**Venae cavae.** The observation that angioplasty is unsuccessful in correcting a dynamic form of baffle obstruction is not surprising; the observation is nonetheless important, because it indicates that in some cases, careful predilation identification of the nature of venous obstruction may predict the success or failure of the procedure.

The results of fixed vena caval baffle obstructions are more gratifying, with resolution of clinical symptoms in all children and an average 75% reduction in the gradient. Six month follow-up in the three survivors has indicated no evidence of gradient recurrence. In two of the children, echocardiographic studies have provided indirect anatomic evidence of persistent improvement, findings that were confirmed in both at follow-up catheterization (table 3).

For angioplasty to succeed in these vena caval obstructions, very large balloons were required, ranging from 6 to 10 times larger than the diameter of the native obstruction. In each case, relatively high dilating pressures (see table 2) were used, although sequential angiograms were not performed after attempted dilations at first low and then high dilating pressures. As a result we cannot be certain how high dilating pressures need to be for a procedure to be successful.

Although the technical details of “caval” angioplasty that appear to contribute to success can be tentatively identified, the anatomic structure being dilated and the mechanism of angioplasty in these cases are less certain. Previous work in animals has suggested that, in arteries, angioplasty succeeds by splitting the intima and extending the tear a variable distance through the media. The medial fibers remain separated and heal in an opened position. However, veins, even those near the heart, have very little media; thus there seems to be little room for a partial venous tear to contribute to an enlarged diameter. Although postoperative vena caval obstructions are not uncommon after surgery for congenital heart disease, the pathologic nature of these lesions has not been precisely identified. In the successfully dilated postoperative baffle obstructions, either pericardium or Dacron was used to fashion the baffle. Neither material has a media that can be partially torn. It is possible but unlikely that angioplasty succeeds in venous obstructions only when there is markedly thickened intima, and it is the intima that partially tears and heals in an open position. It is also possible that thickened intima can be “sieved” through a suture line, allowing for gradient relief. The most likely possibility, however, is that it is atrial myocardial wall that is partially torn, allowing healing in an open position. However, since pathologic data are available for none of the four children with postoperative baffle obstructions, we cannot determine why balloon angioplasty was successful in them.

In summary, we have found that dilation angioplasty of pulmonary venous obstructions is a largely unsuccessful procedure, although the causes for failure vary depending on the nature of the venous obstruction. Postoperative caval or baffle obstructions, in contrast, can be successfully managed with dilation angioplasty, although large balloons are required.

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