

# An Alternative Method for Repair of Partial Anomalous Pulmonary Venous Connection to the Superior Vena Cava

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**ABSTRACT** The surgical management of 15 patients with partial anomalous pulmonary venous connection (PAPVC) to the high superior vena cava (SVC) is described. This new technique redirects the anomalous pulmonary venous flow into the left atrium through the cardiac end of the SVC, transected and oversewn above the anomalous pulmonary vein or veins, by coaptation of the atrial septal defect (or of the surgically created septal defect in patients with an intact atrial septum) to the intracardiac orifice of the SVC. Normal SVC–right atrial flow is reconstituted by atriocavoplasty to the cephalad portion of the transected SVC. A 31-year-old woman with severe pulmonary hypertension died early in the series; this was the only death. Surviving patients enjoy full activity. Except for one symptomatic SVC obstruction due to technical error (since relieved), this technique has achieved total correction of these congenital defects with marked reduction in the undesirable postoperative sequelae often associated with other methods of repair.

Congenital defects in the interatrial septum were described more than a century ago [1, 2]. However, it was not until the emergence of cardiac surgery that serious attention was paid to the anatomy of these anomalies, especially the high-lying sinus venosus defects that more often than not are associated with partial anomalous pulmonary venous connection (PAPVC) to the superior vena cava (SVC) or the right atrium (RA) from one or more of the lobes of the right lung.

Ideal surgical repair demands complete closure of the septal defect with redirection of the anomalous pulmonary venous return to the left atrium without attenuation of pulmonary venous or SVC–RA flow and without injury to the sinoatrial node. That this ideal has not yet been achieved is evident from the large number of varied techniques that have been described [3–9] and from the postoperative follow-up studies that report an undesirable incidence of sinus node dysfunction, SVC or pulmonary venous obstruction, and persistent shunts, mostly left to right but occasionally right to left [10, 11].

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This report describes a corrective surgical technique that has been used since 1967 in patients who had PAPVC to the high SVC with or without an associated atrial septal defect (ASD). Patients who had PAPVC to the low SVC, or the RA, or both during this period were treated by septal transposition [5] or by redirection of the anomalous pulmonary venous flow to the left atrium through the septal defect using a patch applied from within the RA [4]. They have not been included in this report.

## Material and Methods

Between August, 1967, and November, 1983, 15 patients with PAPVC to the high SVC were operated on. *High* is defined as 1 cm or more above the cavoatrial junction. Twelve patients had an associated sinus venosus ASD; 1 of these 12 also had a large foramen ovale. Three patients had intact atrial septa. There were 8 male and 7 female patients. Age at operation ranged from 4 to 52 years (mean, 16 years; median, 12 years). The origin of the anomalous pulmonary veins was from the right upper lobe in 5 patients and from the right upper and middle lobes in 9 patients. In the remaining patient, who had an intact atrial septum, all veins from the right lung coalesced into one large vessel, which emptied into the SVC [12]. Except for 1 patient who had a high SVC connection of a vein from the right upper lobe and a right atrial communication from the middle lobe, all of the central connections of the veins were to the SVC. Seven of these were single and 8 were multiple connections. Three patients also had a persistent left SVC; in 1, there was no communication between the two systems.

## Surgical Technique

In 10 patients the surgical approach used was a right anterolateral thoracotomy through the fourth interspace, and in the remaining 5 a median sternotomy was employed. Repair can be accomplished with equal ease through either incision. The pericardium is opened by a vertical incision parallel and anterior to the right phrenic nerve. The lateral SVC and the posterior RA are inspected to identify anomalous pulmonary veins; bulbous enlargement of the SVC at its atrial junction suggests this. In the case of a sternal approach, the right pleural cavity is entered to ascertain the origin of these veins. Then, the SVC is isolated by sharp dissection and reflection of the pericardium and mediastinal pleura all the way up to the junction with the left innominate vein. Special care must be exercised to avoid injury to the

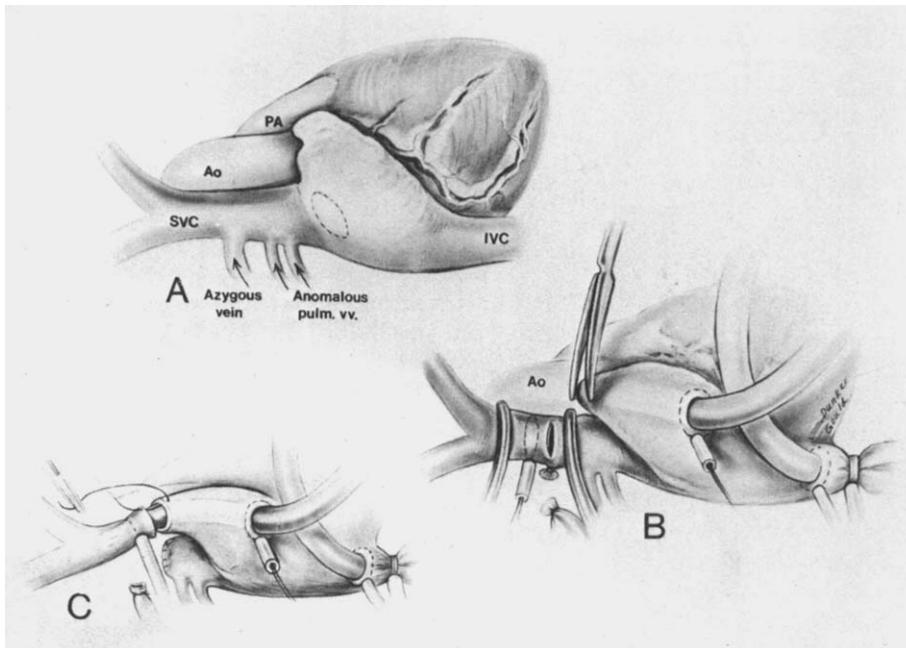


Fig 1. (A) Anomalous pulmonary venous connection to the superior vena cava (SVC) well above the cavoatrial junction. An associated high sinus venosus atrial septal defect is shown by dashed lines. (B) First stage of repair. A superficial pursestring suture is placed around the cephalad SVC, and partial cardiopulmonary bypass is instituted with venous return from the inferior vena cava (IVC). (C) Total cardiopulmonary bypass is established by a catheter introduced through the tip of the right atrial appendage into the cephalad SVC. Caval transection is completed, the stump of the cardiac end of the SVC is oversewn, and anastomosis of the cavoatrial appendage is begun. See text for details. (PA = pulmonary artery; Ao = aorta; vv. = veins.)

closely adherent phrenic nerve. This approach clearly exposes the azygos vein and the anomalous vein or veins (Fig 1A). A pursestring suture of 2-0 Tevdek is superficially tacked to the adventitia around the circumference of the SVC above the azygos vein and the proposed site of anastomosis of the cephalad SVC to the right atrial appendage. The ends of the suture are then passed through a segment of red rubber catheter that serves as a choker. Figure 1B shows the doubly ligated and transected azygos vein. There is some advantage in deferring this step until after cannulation of the upper SVC, as the intact azygos vein serves as countertraction for the maneuver.

Partial cardiopulmonary bypass is instituted after heparinization and cannulation of the ascending aorta and of the inferior vena cava. The cannula for drainage of the SVC is inserted through a pursestring suture placed well down on the lateral wall of the RA and threaded into the appendage. The tip of the appendage is amputated at a level at which the stretched diameter of the appendage approximates that of the cephalad SVC. Complete excision of the trabeculations within the appendage enhances its expandability and reduces the

chance of subsequent adherence or thrombosis. The SVC catheter is then threaded out the appendage. The cava is double-clamped, and the catheter is inserted through a transverse incision in the SVC up to the junction of the innominate vein after release of the upper occluding clamp (see Fig 1B); it is secured by tightening the choker. Total bypass is established and cardioplegia induced. Then, caval transection is completed, the stump of the cardiac end is oversewn with 4-0 suture, and a cavoatrial anastomosis is created using continuous 5-0 suture of the surgeon's choice interrupted in at least two places (Fig 1C).

After completion of the cavoatrial anastomosis, a high right atriotomy exposes the sinus venosus defect (Fig 2A). These defects have a clear-cut, crescent-shaped lower margin that blends anteriorly and posteriorly into the medial cavoatrial junction. There is no discrete upper margin. Repair is accomplished by coaptation of the intracardiac orifice of the SVC to the defect by a continuous 3-0 suture beginning at the anterior angle of the defect and running posteriorly, approximating the free margin of the defect to the atrial wall just beneath its junction with the SVC (Fig 2B). Occasionally it is necessary to enlarge the defect to a size appropriate to that of the SVC. In the 5 patients with an intact septum in this series, high-lying defects were created by excision of an appropriate-sized portion of the septum to achieve the desired repair. The completed procedure is depicted in Figure 2C.

## Results

There was 1 death among the 15 patients who underwent the operation just described. A 31-year-old woman, operated on in 1967, had rapidly progressing symptoms. Cardiac catheterization five months before

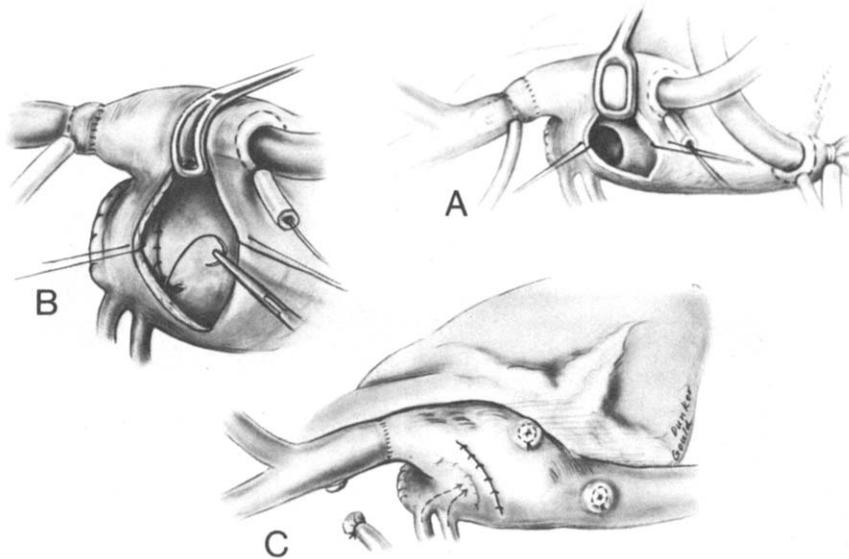


Fig 2. (A) Completed cavoatrial anastomosis. Right atriotomy exposes the sinus venosus defect. In patients with anomalous connection to the superior vena cava (SVC) and intact septa, a high-lying defect is created. (B) The septal defect is coapted to the intracardiac orifice of the SVC by using continuous suture to approximate the inferior margin of the defect to the anterior and right lateral margins of the caval orifice. (C) Completed repair. Anomalous pulmonary venous flow is directed through the stump of the SVC and then through the septal defect into the left atrium, and the left-to-right shunt is eliminated. Flow from the SVC to the right atrium is reconstituted by the cavoatrial anastomosis.

operation demonstrated a 40% left-to-right shunt at the atrial level, no detectable right-to-left shunt, a pulmonary artery pressure of 75/35 mm Hg (mean, 55 mm Hg), an approximated pulmonary vascular resistance of 7 units, and a persistent left SVC. At operation, the pulmonary artery pressure was found to have increased to 95 mm Hg systolic. A 3 cm sinus venosus defect and two anomalous connections (from the right upper and right middle lobes) were repaired as described. Operation was well tolerated, and the early postoperative period was uneventful. However, signs of progressive refractory low output appeared, and the patient died suddenly on the second postoperative day. At autopsy, aside from an uncompromised repair and large pulmonary arteries, the critical finding was grade IV Heath-Edwards changes [13] in the pulmonary arterioles.

Of the surviving 14 patients, 13 have remained asymptomatic. One patient was without symptoms for four months; shortly thereafter, clinically apparent SVC obstruction developed that became complete by eleven months, as confirmed by angiogram of the SVC. At

reoperation one month later, total obstruction at the cavoatrial anastomosis was found secondary to dense connective tissue proliferation between multiple muscular trabeculations on the appendage side of the anastomosis. These were excised at reoperation, and the anastomosis was augmented by pericardial patch angioplasty. Five years postoperatively, the patient had no symptoms. Free cavoatrial flow was documented by angiography.

Follow-up of this group of patients has extended from 7 months to 14 years (mean, 8 years). Postoperative evaluation has included history and physical examination, chest roentgenogram, electrocardiogram, right heart catheterization and angiography, and radionuclide shunt studies. One patient was lost to follow-up 4 years after operation. When last seen, he was asymptomatic and had no abnormal electrocardiographic changes or detectable heart murmur. A chest roentgenogram showed normal pulmonary vascular markings and heart size.

Preoperative and postoperative chest roentgenograms were available for comparison of the 14 surviving patients. A notable reduction in heart size and pulmonary vascular markings occurred in all of them. As might be expected, the degree of improvement tended to have an indirect relationship to the age at operation and a direct relationship to the length of follow-up; children operated on early in the series showed the greatest change.

All patients were invited to undergo postoperative right heart catheterization. Four declined because they were completely asymptomatic and could not be convinced of the value of the study. One, as mentioned, was lost to follow-up; two others have agreed to study at

## Postoperative Catheterization Data

| Patient No., Sex | Age at Operation (yr) | Cath. (mo postop) | Mean Pressure (mm Hg) |     |     |      |      |
|------------------|-----------------------|-------------------|-----------------------|-----|-----|------|------|
|                  |                       |                   | SVC                   | RA  | MPA | RPW  | LPW  |
| 1. F             | 4.5                   | 87                | 5                     | 4   | 14  | 8    | .... |
| 2. F             | 8.5                   | 72                | 12                    | 11  | 15  | 9    | .... |
| 3. F             | 5                     | 55                | 3.5                   | 2   | 12  | 8    | 7    |
| 4. F             | 14                    | 24                | 6                     | 4.5 | 20  | 8    | .... |
| 5. M             | 28                    | 11                | Occl. <sup>a</sup>    | 5   | 15  | .... | .... |
| 6. M             | 9                     | 16                | 11                    | 8   | 24  | .... | .... |
| 7. M             | 5                     | 4                 | 9                     | 7   | 18  | 7    | 9    |

<sup>a</sup>SVC-RA anastomosis was occluded (see text for details).

Cath. = catheterization; SVC = superior vena cava; RA = right atrium; MPA = main pulmonary artery; RPW = right pulmonary wedge; LPW = left pulmonary wedge; Occl. = occluded.

a later time. A summary of the postoperative catheterization data obtained in the remaining 7 patients is shown in the Table. Four patients underwent radionuclide studies. No persistent intracardiac shunts were identified, and other than the caval obstruction already described, no interference with pulmonary venous or caval flow was noted (Figs 3, 4).

Comparative analysis of preoperative and postoperative ECGs was possible in 13 of the 14 patients surviving operation. Three patients had preoperative left-axis deviation of the P wave vector in the frontal plane. One of them had no new electrocardiographic changes until 6 years after operation when atrial flutter developed. Another patient had sick sinus syndrome with predominant junctional rhythm. In the third patient, postoperative tracings were unchanged from those made before operation. In 1 patient who originally had a normal ECG, sick sinus syndrome developed after repair by atriocavoplasty. However, none of the 3 patients with new changes has been symptomatic or required a pacemaker.

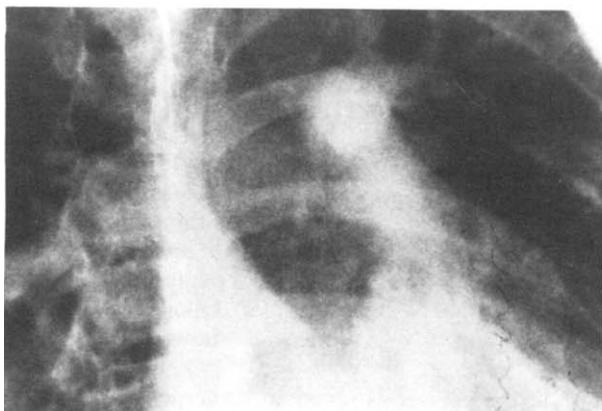


Fig 3. (Patient 2.) Venous cineangiogram from a 14-year-old patient 6 years after operation. The catheter is located at or just above the patulous cavoatrial anastomosis, and there is free flow through the right ventricle into the main pulmonary artery.

## Comment

The first individual in this series may well have been the first to undergo elective repair of PAPVC to the high SVC by the technique we have described here. However, the general plan of this surgical approach had been suggested earlier. In 1958, Lewis [14] contemplated the same technique but did not use it clinically for fear of thrombosis of the venous anastomosis. Also in 1958, Ehrenhaft and colleagues [6] reported on 4 patients in whom they had employed a cavoatrial anastomosis as part of the repair of these anomalies; they used a different technique for closure of the ASD. This method of restoration of cavoatrial flow was reported in 1975 by Pacifico and Kirklin [15] for the dismantling of previously made palliative SVC-right pulmonary artery shunts at the time of total repair of a variety of congenital cardiac anomalies.

On initial inspection, this technique may seem complicated. Actually it is quite simple: it involves one anastomosis, suture closure of the caval stump, and coaptation of the ASD to the orifice of the SVC.

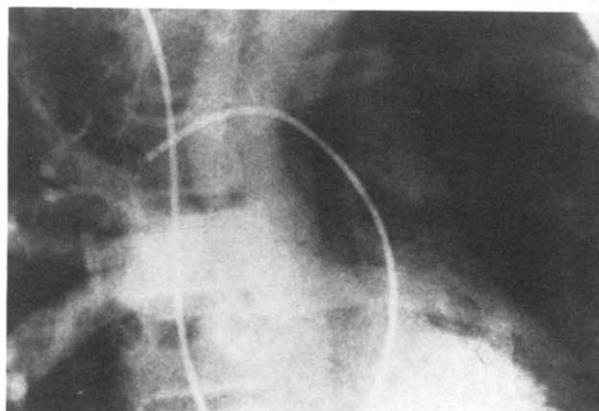


Fig 4. Levophase of right pulmonary cineangiogram from same patient as in Figure 3 shows unobstructed pulmonary venous flow from the right upper and middle lobes into left atrium without residual defect.

The single death was most regrettable. Pressed by progressive, disabling symptoms, the patient's desire for relief, and unwarranted optimism, we undertook the operation. By today's standards, however, the patient's condition was inoperable.

The 28-year-old man who experienced a progressive SVC syndrome postoperatively deserves special comment. It is essential that all of the muscular trabeculae are completely excised from the tip of the right atrial appendage in preparation for anastomosing it to the cephalad portion of the SVC. If this is not done, as occurred in this patient, the intact trabeculae tend to constrict the anastomosis centripetally and increase the chances of subsequent thrombosis. This is especially important in patients in whom the cephalad portion of the right SVC is small because of an associated large communicating persistent left SVC or because of a disproportionate enlargement of the cardiac end of the SVC secondary to large anomalous venous return at this site.

The postoperative course of the 14 surviving patients has been gratifying. All, including the patient relieved of SVC obstruction, have experienced relief of symptoms and have enjoyed unlimited activity. Also supportive of this type of repair for such anomalies is the absence of any residual intracardiac defects or shunts. These results compare favorably with those of other reports, which identify a small but definite number of such undesirable sequelae [16, 17].

Preoperative electrocardiographic findings were similar to those previously reported in that 23% of the patients in this series with sinus venosus ASDs had left-axis deviation of the P wave vector in the frontal plane [18, 19]. This finding in the presence of an ASD should alert one to the possibility that the defect may be of the sinus venosus type, and usually is associated with PAPVC.

Of the 13 patients with available ECGs following this operation, 3 (23%) had rhythm disturbances; these consisted of sick sinus syndrome in 2 and atrial flutter that appeared 6 years after operation in 1. This patient had left-axis deviation of the P wave preoperatively, indicating underlying sinoatrial node dysfunction that may or may not have been made worse by the operation. Fortunately, the resting heart rate has remained normal in all 3 patients and none has required a pacemaker. The incidence of conduction disturbances in this group compares favorably with other series in which as many as 50 to 70% of patients have experienced abnormal cardiac rhythms following other types of repair [11].

## References

1. Rokitansky KF: Die Defecte der Schiedwande des Henzens. Wein, Braumuller, 1875
2. Peacock TB: Diseases, et cetera, of the organs of circulation: I. Malformations of the heart: large aperture in the septum of the auricles: with the foramen ovale closed. *Trans Pathol Soc Lond* 29:43, 1878. Cited by Edwards JE: In Gould SE (ed): *Pathology of the Heart*. Second edition. Springfield, Thomas, 1960, p 269
3. Lewis FJ, Taufic M, Varco RL, Niazi S: The surgical anatomy of atrial septal defects: experiences with repair under direct vision. *Ann Surg* 142:401, 1955.
4. Kirklin JW, Ellis FH Jr, Wood EH: Treatment of anomalous pulmonary venous connections in association with interatrial communications. *Surgery* 39:389, 1956
5. Bahnson HT, Spencer FC, Neill CA: Surgical treatment of thirty-five cases of drainage of pulmonary veins to the right side of the heart. *J Thoracic Surg* 36:777, 1958
6. Ehrenhaft JL, Theilen EO, Lawrence MS: The surgical treatment of partial and total anomalous pulmonary venous connections. *Ann Surg* 148:249, 1958
7. Schuster SR, Gross RE, Colodny AH: Surgical management of anomalous right pulmonary venous drainage to the superior vena cava, associated with superior marginal defect of the atrial septum. *Surgery* 51:805, 1962
8. Puig-Massana M, Murtra M, Revuelta JM: A new technique in the correction of partial anomalous pulmonary venous drainage. *J Thorac Cardiovasc Surg* 64:108, 1972
9. Lewin AN, Zavanella C, Subramanian S: Sinus venosus atrial septal defect associated with partial anomalous pulmonary venous drainage: surgical repair. *Ann Thorac Surg* 26:185, 1978
10. Friedli B, Guerin R, Davignon A, et al: Surgical treatment of partial anomalous pulmonary venous drainage. *Circulation* 45:159, 1972
11. Clark EB, Roland JMA, Varghese PJ, et al: Should the sinus venosus type ASD be closed? A review of the atrial conduction defects and surgical results in twenty-eight children (abstract). *Am J Cardiol* 35:127, 1975
12. Frye RL, Krebs M, Rahimtoola SH, et al: Partial anomalous pulmonary venous connection without atrial septal defect. *Am J Cardiol* 22:242, 1968
13. Heath D, Edwards JE: The pathology of hypertensive pulmonary vascular disease. *Circulation* 18:533, 1958
14. Lewis FJ: High defects of the atrial septum. *J Thorac Surg* 36:1, 1958
15. Pacifico AD, Kirklin JW: Take-down of cava-pulmonary artery anastomosis (Glenn) during repair of congenital cardiac malformations. *J Thorac Cardiovasc Surg* 70:272, 1975
16. Kyger ER III, Frazier OH, Cooley DA, et al: Sinus venosus atrial septal defect: early and late results following closure in 109 patients. *Ann Thorac Surg* 25:44, 1978
17. Trusler GA, Kazenelson G, Freedom RM, et al: Late results following repair of partial anomalous pulmonary venous connection with sinus venosus atrial septal defect. *J Thorac Cardiovasc Surg* 79:776, 1980
18. David JE, Cheitlin MD, Bedynek JL: Sinus venosus atrial septal defect: analysis of fifty cases. *Am Heart J* 85:177, 1973
19. Neal WA, Moller JH, Varco RL, Anderson RC: Operative repair of atrial septal defect without cardiac catheterization. *J Pediatr* 86:189, 1975