Totally anomalous pulmonary venous connection directly to the superior caval vein

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Abstract

Objective: Totally anomalous pulmonary venous connection directly to the superior caval vein is unusual. It is frequently associated with major congenital heart defects such as the syndrome of right isomerism. While improved results have been reported recently for the isolated form, complex cases are still associated with a higher mortality. Methods: We undertook surgical correction in nine patients with direct pulmonary venous connection to the superior caval vein investigated in our institution from 1991 to 1999. In four of these patients, the venous anomaly was an isolated finding, while five patients with isomorphic right atrial appendages had associated cardiac malformations rendering them unsuitable for biventricular repair. In one patient with an isolated form, the venous drainage was obstructed. Two patients with isomerism had previously undergone construction of an aortopulmonary shunt. Other associated surgical procedures in the patients with right isomerism were establishment of bidirectional cavo-pulmonary anastomoses in four cases and banding of the pulmonary trunk in one. Results: There were neither early nor late deaths. Reoperation was needed in one patient because of pulmonary venous stenosis. In the five patients with right isomerism, two later underwent successful creation of the Fontan circulation. Conclusion: It is unusual to find direct drainage of all the pulmonary veins to the superior caval vein. When seen, the venous pathway is only rarely obstructed. For this reason, when associated with right isomerism, an aortopulmonary shunt should be constructed as initial palliation, with later repair of the anomalous pulmonary venous drainage at the time of construction of a bidirectional cavo-pulmonary anastomosis. When using this policy, the surgical results can be as good for the complicated variant as for the isolated form. © 2002 Elsevier Science B.V. All rights reserved.

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1. Introduction

In most of a large series describing totally anomalous pulmonary venous connection, the site of drainage is most frequently supracardiac [1–3]. The connection is usually to a left vertical vein, which then drains into the brachiocephalic vein. Less often, the drainage is directly to the superior caval vein and rarely to the caval vein via the azygos vein. These patterns are frequently seen in association with complex intra-cardiac malformations, rendering the patients unsuitable for biventricular repair. This setting mandates perfect preoperative evaluation, especially for those patients who are candidates for a future Fontan operation. In this report, we review our experience in the treatment with such patients, paying particular attention to our surgical management of those with complex associated malformations.

2. Materials and methods

During the period from January 1991 to October 1999, we undertook surgical correction in 100 patients with totally anomalous pulmonary venous connection (Table 1). In nine of these, the drainage was directly to the right superior caval vein and they were selected for this study. Their age at operation ranged from 3 days to 8 years, with a mean age of 30 months. One patient, without other complicating features, had obstruction of the anomalous venous connection (Fig. 1). Six patients underwent preoperative cardiac catheterisation.

In six patients, the four pulmonary veins were situated behind and below the right pulmonary artery. They drained into a common venous channel, which extended superiorly to enter the superior caval vein posteriorly to the cavo-atrial junction, and below the azygos vein. In two patients, the left and the right pulmonary veins entered separately at each side of the superior caval vein. In the last patient, the right pulmonary veins entered directly to the back of the caval
vein, while the left pulmonary veins, having joined to form a large horizontal collecting vein, passed behind the heart to terminate in the superior caval vein (SVC) (Fig. 2).

Four patients had no other complicating lesion, apart from an inter-atrial communication within the oval fossa. It was in one of these patients that the venous connection was obstructed. The other five patients, all with right isomerism, presented with major associated cardiac malformations, making them unsuitable for biventricular repair (Table 2). All five of these patients underwent preoperative cardiac catheterisation.

Three patients had undergone previous surgical procedures. Systemic-to-pulmonary shunts had been constructed in two patients with right isomerism. An atrial septectomy, combined with an attempt to repair the venous anomaly, had been undertaken at another institution in one of the patients without associated malformations. This patient underwent tracheostomy after the initial surgery.

2.1. Operative techniques

The anomaly was repaired in six patients by making a posterior anastomosis between the roof of the left atrium, or the left-sided atrium in those with right isomerism, and the pulmonary venous confluence. The confluence itself was mobilised extensively and transected from the caval vein, the venous tissue being used as a gusset to enlarge the anastomosis. In three patients, in order to achieve complete mobilisation of the pulmonary veins, the superior caval vein was transected just below the azygos vein.

Additional surgical procedures were needed in six patients. Bidirectional cavo-pulmonary anastomoses were constructed in two and the pulmonary trunk was banded in one. In another patient, the superior caval vein was reconnected to the cavo-atrial junction using a 24 mm Gore-tex tube. This patient also needed plication of the right hemidiaphragm. The atrial septal defect was closed with a Gore-tex patch in two patients. In two patients, the superior caval vein was transected above the pulmonary veins and used to construct a bidirectional cavo-pulmonary anastomosis. The cavo-atrial junction was then closed with a Gore-tex patch above the initial site of drainage of the pulmonary veins. In the last patient, with an isolated anomalous connection, the superior caval vein was transected above the pulmonary veins. The atrial septal defect was then closed with a Gore-tex patch, rerouting the pulmonary venous flow

| Sites of drainage in 100 cases of totally anomalous pulmonary venous connection |
|---------------------------------|---------|
| Supracardiac                    | 45      |
| Direct to superior caval vein   | 9       |
| Cardiac                         | 24      |
| Infracardiac                    | 21      |
| Mixed                           | 10      |
| Total                           | 100     |

Table 2

Associated cardiac malformation in patients with right isomerism

| DIRV, CA, common AVV, severe PS       | 2       |
| DIRV, CA, common AVV                  | 1       |
| DILV, right-sided heart, common AVV, LSCV, moderate PS | 1 |
| DIRV, mitral atresia, DORV, moderate PS | 1 |
| Total                                | 5       |

CA, common atrium; AVV, atrio-ventricular valve; PS, pulmonary stenosis; LSCV, persistent left superior caval vein; DORV, double outlet right ventricle; DIRV, double inlet right ventricle; DILV, double inlet left ventricle.
towards the left atrium through the defect. The superior caval vein was then anastomosed with the right atrial appendage.

3. Results

There were no deaths, either within 30 days or prior to discharge from hospital. In one patient undergoing repair of the anomalous venous connection and construction of a bidirectional cavo-pulmonary anastomosis, a very small right superior pulmonary vein remained connected to the superior caval vein. This created non-physiological perfusion of the right upper lobe, which became virtually useless. This patient was treated with heparin infusion and oral anticoagulants.

Over a mean follow-up of 37 months (1–78 months), there have been no late deaths. Reoperation was needed in one patient 4 months after the initial procedure because of pulmonary venous stenosis. This patient underwent Gore-tex patch enlargement of the previous anastomosis site. In the group of five patients with right isomerism, two patients subsequently underwent a successful Fontan operation. These were performed 12 and 62 months, respectively, after repair of the anomalous venous connection and construction of the bidirectional cavo-pulmonary anastomosis. The connection between the inferior caval vein and the pulmonary arteries was established with an extracardiac Gore-tex conduit in one patient and using an intra-extracardiac fenestrated Gore-tex conduit in the other [4].

4. Discussion

The supracardiac variant is not only the commonest, but also the most variable form of totally anomalous pulmonary venous connection. Drainage can be to a left vertical vein and thence to the brachiocephalic vein, less often directly to the right superior caval vein at its junction with the right atrium, and rarely to the caval vein via the azygos vein. Obstruction to venous return is known to occur in about two-fifths of all the cases with supracardiac drainage [1–3] but is less frequent when the drainage is directly to the superior caval vein. In those with obstruction, the anomalous confluence usually passes posteriorly to the right pulmonary artery, typically being compressed between it and the right main bronchus.

A perfect strategy for surgical repair is mandatory to produce optimal immediate and late results, especially for those patients in whom associated major cardiac malformations render them unsuitable for biventricular repair, such as those with right isomerism. It is the supracardiac variant of totally anomalous pulmonary connection, which is seen most commonly in patients with right isomerism, excluding those in which the pulmonary veins join anomalously to the atrial chambers [5,6]. When there are no associated cardiac anomalies, surgical management is now known to be associated with very low perioperative mortality [2,3,7]. This is not the case in the setting of complex associated malformations. In the complicated cases, particularly when there is reduced flow of blood to the lungs, mortality is close to 50% [8,9]. A higher mortality is also reported when the surgical correction of the anomalous venous connection is associated with banding of the pulmonary trunk [9].

The severity of the association is confirmed by an unacceptably high mortality in the staging procedures towards the Fontan operation [10]. Because of this, heart transplantation has been proposed by some for these complex patients [10]. It is the association of potentially abnormal pulmonary inflow and outflow, which makes the primary palliation particularly difficult. The pulmonary outflow tract is obstructed in the majority of patients with right isomerism [5]. Creation of a stable source of the pulmonary flow by a shunt, therefore, should be the initial goal.

Increased pulmonary flow following construction of the shunt, however, may unmask relative pulmonary venous obstruction, with the need for concomitant repair of the anomalous pulmonary venous connection. Results of such concomitant repair as part of the initial palliation are also discouraging [5,8,9]. In patients with unobstructive pulmonary venous drainage, therefore, management is by construction of an aortopulmonary shunt alone. The mortality for this procedure seems to be acceptable [10]. This permits later repair of the anomalous venous connection at the time of the bidirectional cavo-pulmonary anastomosis as staging towards the Fontan procedure.

Of the five patients in our series with right isomerism, two had a primary shunt and secondary construction of a bidirectional cavo-pulmonary anastomosis plus repair of the anomalous connection. Both of these patients successfully underwent a subsequent Fontan operation.

We performed primary repair along with construction of a bidirectional cavo-pulmonary anastomosis in two of the remaining patients, with repair together with banding of the pulmonary trunk in the other. All these patients are awaiting completion of the Fontan circulation (Table 3).

It is crucial to identify precisely the anatomy of the pulmonary veins preoperatively. In one of our patients, an unexpected very high insertion of the right superior pulmonary vein was overlooked, with the vein remaining after surgery to drain into the bidirectional cavo-pulmonary anastomosis. This drainage resulted in equalisation of pressures in the corresponding pulmonary artery and vein, making the lobe useless and at high risk for cessation of flow, thrombosis, and pulmonary infarction. The patient was aggressively treated with anticoagulants [11].

Several techniques have been proposed for repair of anomalous supracardiac drainage [3,12,13]. We prefer the superior approach because it provides excellent exposure of the common vein and the roof of the left atrium in the same plane, reducing the risk of kinking at the site of anastomosis. With such good exposure, coupled with the elevated mean age of our patients, we did not need to divide the ascending
of the enlarge the anastomosis. This technique permits elimination the superior caval vein. The venous tissue is then used to

References

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