Complications following closure of atrial septal defects of the inferior vena caval type

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An atrial septal defect of the inferior vena caval type presents complicated anatomy to the surgeon at operation. Four cases are presented illustrating two complications of repair. Incomplete closure or reopening due to disruption of the suture line may be prevented by accurate identification of the anatomy and a patch closure. Two cases of inadvertent diversion of the inferior vena cava into the left atrium are presented and the methods of diagnosis and techniques of repair are discussed. Mitral regurgitation due to ballooning of the posterior cusp in systole was an associated lesion in two cases of atrial septal defect of this type.

The secundum type atrial septal defect most commonly needs closure in young asymptomatic patients, so that any surgical complication is particularly disconcerting. The inferior vena caval type of defect presents more of a technical problem than the fossa ovalis defect, having no inferior septal margin and sitting astride the orifice of the inferior vena cava with intimate relationships to the valve of the inferior vena cava and the right inferior pulmonary vein orifice.

These defects are liable to be incompletely closed at the lower end, or when sutured under tension the repair may disrupt postoperatively. Inaccurate closure may also produce a diversion of the inferior vena caval blood flow into the left atrium (Fig. 1). Bedford et al. (1957) and Björk et al. (1958) described the presentation of this complication during or immediately after operation when hypothermia had been used. More commonly, as described by Effler and Groves (1961) and Mustard, Firor, and Kidd (1964), there is an apparently uncomplicated immediate postoperative course with satisfactory, even dramatic, reduction in heart size and pulmonary vascularity, only to be followed by the insidious onset of cyanosis, dyspnoea with effort, and eventually finger clubbing and polycythaemia.

We report and discuss the techniques we have used to deal with these problems in four clinical cases.

CASE HISTORIES

CASE 1  J.H., a 26-year-old woman, underwent repair of an asymptomatic atrial septal defect under hypothermia at another hospital in January 1964. The secundum defect of inferior vena caval type measured 6.5 x 3 cm, being an almost total defect of the lower interatrial septum. The immediate postoperative course was uneventful with marked reduction in heart size on the chest radiograph, but over the next few months she complained of dyspnoea on walking and climbing stairs and thought on several occasions that she was cyanosed.

In October 1964, right heart catheterization and angiocardiography via the superior vena cava were performed but gave no useful information except that the systemic arterial oxygen saturation was 87%. Over the next three years she consistently complained of increasing cyanosis and dyspnoea while walking or climbing stairs but she was able to carry on her favourite recreation of bell-ringing in the local church without difficulty, an activity which entailed exercise of the upper limbs only.

In August 1967, another right heart catheterization was performed and on this occasion the catheter was passed from the right long saphenous vein up the inferior vena cava. The catheter passed into both right and left atria and an angiocardiogram showed that...
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A differential exercise test revealed a significant fall in systemic oxygen saturation to 64.5% with leg exercise but no significant change with arm exercise (Table).

In January 1968 she was admitted to the National Heart Hospital for corrective surgery. On examination there was mild cyanosis at rest and mild clubbing of the fingers. A short systolic murmur grade I/IV along the left sternal edge was the only other abnormal physical finding. Haemoglobin was 18.4 g%, PCV 57%, and total WCC 3,800/mm³. A chest radiograph revealed a normal sized heart with normal pulmonary vascularity.

The heart was exposed by median sternotomy, and palpation via the right atrial appendage failed to reveal any communication with the inferior vena cava but the suture line in the septum was easily felt. The dye passed from the inferior vena cava into the left atrium (Fig. 2).

FIG. 2. Case 1. Angiocardiogram. The catheter has been passed up the inferior vena cava and its tip lies at the inferior cavo-atrial junction. An injection of contrast medium has filled the left atrium with reflux into a hepatic vein and the right inferior pulmonary vein.

<table>
<thead>
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<td>PREOPERATIVE SYSTEMIC O₂ SATURATION ON DIFFERENTIAL EXERCISE: CASE 1</td>
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<tr>
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<th>O₂ Content (ml/l)</th>
<th>% Saturation</th>
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<tr>
<td>Rest</td>
<td>136</td>
<td>84.5</td>
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<tr>
<td>Leg exercise</td>
<td>95</td>
<td>64.5</td>
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<tr>
<td>5 minute rest</td>
<td>129</td>
<td>82</td>
</tr>
<tr>
<td>Arm exercise</td>
<td>124</td>
<td>79</td>
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<tr>
<td>10 minute rest</td>
<td>125</td>
<td>80</td>
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A second graft (2) is used to enlarge this area; (c) graft (2) in position, completing the repair.

FIG. 3. Cannulation of the inferior vena cava using the saphenofemoral junction: (a) line of incision in stump of saphenous vein; (b) cannula in position. In practice the cannula would be larger and would fill the femoral vein; (c) the opened-out stump of the saphenous vein can be used as a pedicled patch to close the cannulation site and so avoid narrowing the femoral vein.

FIG. 4. Reconstruction of atrial septum: (a) the inferior vena cava has been mobilized and occluded by a clamp. The previous suture line in the septum (stippled) has been reopened and pericardial graft (1) used to reconstruct the lower end of the septum; (b) the broken line indicates how closure by direct suture produces narrowing at the inferior cavo-atrial junction. A second graft (2) is used to enlarge this area; (c) graft (2) in position, completing the repair.
inferior vena cava was cannulated via the femoral vein (Fig. 3) and the superior vena cava was cannulated via the right atrial appendage, and arterial return was into the ascending aorta. Cardiopulmonary bypass was instituted with cooling to 31.5°C and the inferior vena cava was carefully dissected free down to the diaphragm and occluded by a clamp instead of a snare, thus avoiding distortion of the cava. After clamping the aorta, the right atrium was opened longitudinally in the line of the previous incision. The atrial septum was incised parallel to the previous suture line which had included a prominent valve of the inferior vena cava, diverting the inferior vena caval blood into the left atrium. There was not sufficient septal tissue to reconstruct the septum in its correct anatomical plane so a Dacron patch was sutured in so that it redirected the inferior vena caval blood forward into the right atrium and closed the septal defect (Fig. 4a). Some difficulty was experienced in placing this patch accurately because the inferior pulmonary vein lay close to the line of closure of the septum. The forward angulation of the Dacron patch produced narrowing of the inferior vena cava/right atrial junction which had to be enlarged with a gusset of free pericardium (Fig. 4). There were no complications. She was given anticoagulants for six months to prevent any thrombus forming on the Dacron patch until endothelialization had occurred.

Two years postoperatively she was pink with a normal exercise tolerance.

CASE 2 G.P., an 18-year-old boy, underwent repair of an asymptomatic secundum type atrial septal defect under cardiopulmonary bypass at another hospital in February 1962. The postoperative course was uncomplicated.

Over the next two years faint central cyanosis was noticed on exercise, gradually increasing in severity and eventually becoming established at rest. There was only mild dyspnoea on walking up hills and stairs.

Examination revealed moderate central cyanosis with grade 2–3 clubbing of the fingers. There was sinus rhythm with a blood pressure of 130/110 mmHg and no evidence of cardiac failure. The heart was quiet on palpation and the pulmonary component of the second heart sound moved normally with respiration; there were no murmurs. A chest radiograph revealed a normal sized heart. The lung fields were slightly plethoric, but the main pulmonary artery segment was not prominent. Haemoglobin was 22.4 g%. The electrocardiogram was within normal limits.

In July 1970 cardiac catheterization revealed a systemic arterial oxygen saturation of 80% with normal right and left heart pressures. A catheter passed from the inferior vena cava easily into the left atrium and into all pulmonary veins except the inferior left orifice. An inferior vena cavogram demonstrated the inferior vena cava entering the left atrium.

He underwent open heart surgery at the National Heart Hospital in January 1971. Palpation through the right atrial appendage revealed a small low atrial septal defect which was too small to accept a cannula. This was the only opening into the inferior vena cava. The right femoral vein was exposed and cannulated as described previously and the superior vena cava was cannulated via the right atrial appendage. The ascending aorta was cannulated for arterial return and the patient was placed on normothermic cardiopulmonary bypass. The inferior vena cava was dissected free down to the diaphragm and cross-clamped carefully to prevent distortion. With the aorta cross-clamped the right atrium was opened and the atrial septum incised upwards from the small residual septal defect along the line of the original closure. An elliptical pericardial graft was then sutured into position so as to deflect the inferior vena caval return forward into the right atrium and to close the interatrial septum (Fig. 4). It was necessary to enlarge the inferior vena cava/right atrial junction with a second pericardial graft at the lower end of the incision in the right atrium (Fig. 4).

There were no postoperative complications. Two months postoperatively he was a normal colour and the finger clubbing was regressing.

CASE 3 Y.H., a 35-year-old woman, underwent closure of a large secundum type atrial septal defect by direct suture with cardiopulmonary bypass in 1966 at another hospital. Previously her only symptom had been tiredness after moderate exertion.

Postoperatively, although initially improved, she soon complained of dyspnoea with effort again and the heart failed to decrease in size. On examination there was sinus rhythm with a normal blood pressure and a predominant left ventricle. There was fixed splitting of the second heart sound, a pansystolic apical systolic murmur (grade II/VI), and a third heart sound. A chest radiograph revealed moderate cardiac enlargement and a prominent pulmonary artery with pulmonary plethora. An electrocardiogram showed sinus rhythm with a right axis and right ventricular hypertrophy.

Cardiac catheterization confirmed the presence of a residual atrial septal defect with a pulmonary to systemic flow ratio of 3 to 1 and mild to moderate mitral regurgitation on left ventricular cineangiography.

In March 1971, the right atrium was explored with cardiopulmonary bypass and a large residual septal defect of the inferior vena caval type was found. The original suture line had torn away, leading to complete recurrence of the defect. The mitral valve appeared normal on inspection and there had been no obvious mitral regurgitation on palpation via the right atrial appendage before bypass.

The defect was accurately closed with a Dacron patch. Postoperatively she received anticoagulants for three months. Four months postoperatively the heart has decreased in size and there has been a reduction in pulmonary plethora, but an apical pansystolic murmur persists with a little prominence of the left
atrium. It is thought that she has mitral regurgitation due to prolapse of the posterior mitral cusp.

CASE 4 P.O., a 25-year-old woman, underwent repair of an asymptomatic secundum type atrial septal defect at another hospital at the age of 15 years under hypothermia. A large secundum defect low in the atrial septum was described and the lower margin of the defect merged with the valve of the inferior vena cava. A direct suture closure was performed but was incomplete inferiorly because of possible stenosis of the orifice of the inferior vena cava.

On admission to the National Heart Hospital there were signs of a left-to-right shunt at atrial level and a grade II/IV pansystolic murmur at the apex with a slightly prominent left ventricle. A further cardiac catheterization revealed a pulmonary systemic flow of 3:1 with mild elevation of the pulmonary artery pressure. A left ventricular angiocardiogram showed an abnormal mitral valve with ballooning of the posterior cusp accompanied by mild mitral regurgitation.

In June 1970, under normothermic cardiopulmonary bypass, the right atrium was explored. Before institution of bypass, palpation via the right atrial appendage revealed a low atrial septal defect without a lower margin and trivial mitral regurgitation without mitral stenosis. On opening the right atrium there was a residual inferior vena caval type septal defect sitting astride the orifice of the inferior vena cava. The mitral valve appeared normal except for a redundancy of the centre of the posterior cusp with normal looking chordae. With a dry field using ischaemic arrest the anatomy of the septal defect was carefully identified, and it was closed with a patch of autogenous pericardium.

After bypass the left atrial pressure was 9 mmHg with excellent cardiac function. Six months after surgery she was asymptomatic and the heart was considerably smaller with reduction in pulmonary vascularity. There was still a soft pansystolic murmur at the apex.

DISCUSSION

Rokitansky (1875) first described the anatomy of the atrial septal defect lying astride the orifice of the inferior vena cava, the defect having no lower septal margin, and with the opening of the inferior vena cava directed partly into the left atrium.

Diversion of the inferior vena cava into the left atrium complicating closure of these defects was first described by Bedford et al. (1957) when three cases occurred in 61 closures of atrial septal defect by direct suture using circulatory arrest under mild (30°C) hypothermia by surface cooling. Although there have been other reports in the literature (Björk et al., 1958; Clause et al., 1962; Effler and Groves, 1961; Mustard et al., 1964; Osawa, 1968; Staple, Ferguson, and Parker, 1966), it is known that many other unreported cases have occurred. All authors have emphasized that if the inferior vena caval valve is mistaken for the lower margin of the defect and included in the line of suture closure, then this complication will occur. Although this is more likely under the hurried conditions when mild hypothermia is used, the presence of a venous cannula in the inferior vena cava does not guarantee that this accident cannot happen, as is shown by our second case. Distortion of the inferior vena caval orifice by a snare may also lead to inaccurate placement of sutures (Effler and Groves, 1961), and failure to appreciate the relationships of the lower edge of the atrial septum to the caval orifice may lead to the same complication.

Because the anatomy is more complex, the inferior vena caval type of defect tends to be incompletely closed more often than other types of atrial septal defect, leaving a residual left-to-right shunt. In addition, when a direct suture closure is used, tension is created between the relatively fixed suture line in the septum and the mobile tricuspid valve annulus. This predisposes to partial or complete breakdown of the repair, particularly if there is little or no strong tissue along the anterior or medial margin of the defect. For these reasons it is our practice to close this type of defect with a patch, preferably of autogenous pericardium, inserted in a dry field after accurate identification of the anatomy. The use of a patch prevents tension at the suture line and allows for the difference in plane between the atrial septum and the posterior margin of the inferior vena caval orifice.

The occurrence of an inadvertent diversion of the inferior vena cava into the left atrium may present on the operating table or in the immediate postoperative period with severe cyanosis or hypoxia-induced arrhythmias, as in early reports by Bedford et al. (1957) and Björk et al. (1958). In these cases there was probably total diversion of inferior vena caval flow into the left heart. Both authors reported successful resuscitation of patients by immediate reoperation under hypothermia, in one case preceded by emergency angiocardiography, but there was a residual right-to-left shunt in one case and there were several fatalities. Successful total repair requires a careful reappraisal of the anatomy under ideal conditions, but if facilities are less than ideal, complete reopening of the septal defect is life saving.

Frequently the diversion is not complete initially, particularly when the inferior vena cava has been cannulated for extracorporeal circulation, a
small residual defect remaining. The late presentation of this syndrome in our two cases was classical in that there were no immediate postoperative complications. Gradually, some months later, dyspnoea and cyanosis on exercise involving the lower limbs but not the arms became manifest, and still later cyanosis at rest. Presumably this followed complete or almost total septal closure due to fibrosis and contracture of the margins of the residual defect. Due to abolition of the left-to-right shunt, serial chest radiographs classically demonstrate a dramatic decrease in heart size and pulmonary vascularity with shrinkage of the right atrium. The diagnosis can be confirmed by right heart catheterization via the femoral vein and inferior vena cava and may be missed if the superior vena caval route is used. Systemic arterial blood samples during exercise of the upper and lower limbs demonstrated the differential oxygen saturation in case 1 (Table). Injection of contrast medium via an inferior vena caval catheter and angiocardiology in the oblique plane will clearly show the abnormality (Fig. 2).

Repair of this abnormality requires a carefully planned approach with a dry undistorted field to allow accurate identification of the anatomy. We have used a median sternotomy for exposure and normothermic cardiopulmonary bypass with a period of ischaemic arrest. No reliance should be placed on finding a residual opening between the right atrium and inferior vena cava for cannulation and the inferior vena cava should be cannulated peripherally via the femoral vein. We have preferred to use the saphenofemoral junction as a cannulation site and use the saphenous vein stump in the vein closure (Fig. 3). The inferior vena cava above the diaphragm should be carefully dissected free before or soon after starting bypass and occluded by a clamp at the level of the diaphragm as a conventional snare causes distortion of the inferior cavo-atrial junction and makes the repair more difficult.

The plane of the incorrectly closed septum lies well forward and to the right so that the graft used in the repair after the septum has been re-opened is angled forwards. This tends to narrow the inferior cavo-atrial junction, and in both our cases a gusset was required to enlarge this junction and prevent stenosis (Fig. 4). The septal graft may be of autogenous pericardium or Dacron fabric, and if the latter is used consideration should be given to using a short period of anticoagulation postoperatively to prevent possible thrombosis on the patch before endothelialization occurs.

The occurrence of mild mitral regurgitation in cases 3 and 4 due to ballooning of the posterior mitral cusp in systole was probably not due to chance as the association of this lesion and atrial septal defect has been described by McDonald et al. (1970). Both patients have residual pansystolic apical murmurs and the subsequent fate of these valves is unknown, but there has been a gratifying decrease in heart size and symptomatic improvement in both cases since closure of the atrial septal defect.

REFERENCES