A boy presented with Alagille syndrome and recurrent coarctation of the descending thoracic aorta. Initially, he underwent resection of the coarctation segment and end-to-end anastomosis. Following recurrent coarctation an extra-anatomic bypass procedure (a conduit was interposed between the ascending and descending thoracic aorta) was performed. He recently presented with stenosis at the anastomotic site between the extra-anatomic conduit and ascending aorta. Tissue overgrowth at the anastomotic site was responsible for the ‘recoarctation’. This was surgically relieved. Patients with Alagille syndrome have vascular lesions affecting multiple organs. The management of these patients is developing. We highlight the need to tailor-make treatment to the requirements of the individual, the need for close follow-up, and the importance of long-term management of these patients.

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1. Introduction

Recurrent coarctation is not uncommon and is dependant on age at operation, anatomy of lesion, length of abnormal tissue resected and type of surgical procedure performed [1].

We present a case with Alagille syndrome (AS) and recurrent coarctation. AS is an autosomal dominant multisystemic disease that can affect the aorta and pulmonary arteries [2]. The condition causes vascular dysplasia and may be responsible for the ‘re-coarctation’.

2. Case history

A two-year-old boy presented with exertional dyspnoea, reduced lower limb pulses, proximal hypertension and a heart murmur. Electrocardiogram showed normal sinus rhythm and biventricular hypertrophy. Echocardiogram demonstrated isolated coarctation of the aortic isthmus. Cardiac catheterisation and angiography demonstrated a long segment coarctation with a 90 mmHg pressure gradient. The transverse aortic arch was well developed and the descending thoracic aorta was small. The right subclavian artery had an abnormal origin, it arose immediately distal to the left subclavian artery and passed behind the oesophagus. Both branch pulmonary arteries had mild distal stenoses without any associated pressure gradient.

Through a left lateral thoracotomy the isthmic coarctation was identified. A 14-mm polytetrafluoroethylene (PTFE) shunt was created to channel blood from the aortic arch to the descending thoracic aorta. The aortic arch and proximal descending aorta were mobilised. A 15-mm segment of narrowed isthmus was resected. The descending thoracic aorta was pulled superiorly and directly anastomosed to the underside of the aortic arch. The shunt was removed. Recovery was uneventful with no residual gradient.

One year later there was clinical evidence of recurrence of the coarctation and the echocardiogram showed a residual narrowing at the site of the previous anastomosis with a mean gradient of 20 mmHg. Percutaneous balloon dilatation of the stenosis was attempted but was unsuccessful.

The patient remained clinically well, but at four-years of age he was found to have stenosis of the main and left branch pulmonary arteries. Lung scans showed reduced left lung perfusion consistent with the left pulmonary artery stenosis. The additional presence of impaired liver function, butterfly-shaped deformity of thoracic vertebrae and abnormal facial features prompted the diagnosis of AS [2]. Defects in the Jagged-1 gene [3] confirmed the diagnosis of AS in the patient, his twin brother and their father.

The aortic coarctation became more severe and at five-years of age he had a second cardiac operation. Through a right thoracotomy, cardiopulmonary bypass (CPB) was established between the right atrium and the right femoral artery. A 14-mm PTFE conduit was placed between the ascending and the descending thoracic aorta (Fig. 1). Recovery was uneventful and no pressure gradient across the coarctation was demonstrated by direct pressure measurement or seen on Doppler echocardiogram.

Two years later, temporary left arm weakness, left facial nerve palsy and slurred speech developed acutely. Stenoses of both carotids and the intracerebral arteries were demonstrated on magnetic resonance imaging. He underwent
bilateral carotid arterioplasty and made a complete recovery.

A Doppler gradient of 35 mmHg across the coarctation was found at routine cardiac follow-up. CT scan (Fig. 2) demonstrated a stenosis at the anastomosis between the conduit with the ascending aorta.

A third cardiac operation was performed at eleven-years of age through a median sternotomy. CPB was established between the right atrium and ascending aorta. Clamps were placed across the conduit and partially across the ascending aorta. An overgrowth of fibrous tissue with a 2-mm central lumen lay within the proximal conduit and extended into the aorta. This was resected and the conduit was reanastomosed to the ascending aorta. He recovered quickly and there was no evidence of coarctation at four month follow-up.

3. Discussion

AS affects the liver, heart, vascular tree, eyes, face and skeleton [2]. Branch pulmonary arteries stenosis is common affecting 76% of subjects [3], but does not appear to be progressive [4]. With this in mind, we did not attempt to intervene when the branch pulmonary artery stenosis was identified. Left-sided cardiovascular anomalies affect mainly the aorta and occur in 11% of patients [4].

Intracranial bleeding increases morbidity and mortality in AS patients [3]. CPB could have increased this risk and refashioning of the anastomosis could have been performed without it. In this case the benefits of maintaining cerebral and peripheral perfusion pressures with CPB-support were felt to have out-weighed the risks of cerebral haemorrhage with systemic heparization.

The initial coarctation repair with resection and end-to-end anastomosis produces good long-term results [5]. It was felt that the abnormal right subclavian artery origin may have an unknown effect if the left subclavian artery was used as a flap, especially as this approach would also put the left arm at risk of ischaemia. AS was diagnosed when the re-coarctation occurred. It was considered that another attempt at coarctation repair would prove difficult and that the vascular dysplasia associated with AS may be responsible for the re-coarctation. Therefore, the right chest approach and extra-anatomic conduit was chosen. The tissue overgrowth at the anastomotic site that followed this procedure may again have been due to the underlying AS pathology (underlining our previous concerns). We also suggest that the vascular dysplasia of AS is responsible for the vascular tissue being unsuitable in that it does not lend itself to conventional healing.

The extra-anatomic approach to coarctation repair has low associated morbidity and mortality and long-term results have been good in adults [5, 6]. Coarctation is more likely to recur when the initial operation is performed at a young age, when the coarctation segment is long or when synthetic materials are used for tissue reconstruction [1]. The underlying vascular condition appeared to cause recurrence in this case. The long-term results of the extra-anatomic technique in childhood have not been reported. As the child grows the relatively fixed conduit would cause concern. The overgrowth of tissue that resulted in the anastomotic stenosis appears to be a new complication and was easily dealt with in this case.

Technical factors during the anastomosis may play a roll in recoarctation. Additional techniques that may be used include the use of patches during the repair and endovascular balloon dilatation. Homograft or biological tissue patches are preferable as synthetic patches are associated with late aneurysm formation [7].

Apart from highlighting the unusual presentation, we suggest that the presence of vascular dysplastic conditions may affect the outcome of any of the appropriate surgical procedures used for treating coarctation; and that these patients need close follow-up. This was, and continues to be, a difficult case to manage. The need to tailor-make the surgical management to the specific congenital cardiac anatomy and the associated medical condition is also highlighted.

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References


