

## Editorial

# Stenting of systemic venous pathways after atrial repair for complete transposition

Atrial repair (Mustard and Senning operations) for complete transposition has been replaced in most paediatric cardiology centres by the arterial switch operation, which is performed in the first few days or weeks of life.<sup>1</sup> Arterial switch avoids many of the major complications of atrial repair: sinus node damage results in loss of sinus rhythm, atrial arrhythmias, bradycardias, and the need for antiarrhythmic medication and pacemaker implantation.<sup>2-3</sup> Systemic (morphological right) ventricular dysfunction and failure, when it occurs, requires either heart transplantation or takedown of the original repair and conversion to an arterial switch operation.<sup>4-5</sup> Late sudden death is not always related to these complications and is unpredictable.<sup>2-3,6</sup> Systemic and pulmonary venous pathway obstruction can cause significant morbidity and mortality as does repeat surgery.<sup>7</sup> Balloon dilatation and, recently, stent implantation have been used to avoid repeat surgery for pathway obstruction.<sup>8-11</sup> A trend towards a "double switch" (Senning or Mustard operation *and* arterial switch or Rastelli procedure) in patients with congenitally corrected transposition (atrioventricular and ventriculoarterial discordance) and the occasional patient with complete transposition who is unsuitable for an arterial switch operation, suggest that these problems will not be restricted to patients from an earlier surgical era.<sup>12</sup>

Severe obstruction or occlusion of the systemic venous pathways leads to systemic venous hypertension, which, while occasionally asymptomatic, may present with superior vena caval syndrome, peripheral oedema, and protein losing enteropathy.<sup>13-15</sup> The incidence varies considerably and is probably less common after the Senning operation in which prosthetic material is avoided when forming the interatrial baffle, or after the Mustard operation if Dacron is avoided.<sup>7,16</sup> Some reports, however, suggest a similar incidence of complications for the Mustard and Senning operations.<sup>17</sup> There are no published reports of stent implantation after the Senning operation although this has been encountered and stent implantation is performed in our unit.

Complete occlusion of the superior vena cava pathway after atrial repair may be well tolerated because of collateral formation to the inferior vena cava, and is often only clinically evident with co-existent narrowing of the inferior caval pathway. Near occlusions may be at risk of progression to complete occlusion, but the natural history of mild to moderate asymptomatic systemic venous stenoses is not clear. Thus, in asymptomatic patients, it is uncertain at what degree of stenosis intervention should be performed. The importance of mild gradients across the systemic venous pathways (arbitrarily < 3 mm Hg) is also unknown, while gradients of 10–15 mm Hg are reported in patients with protein losing enteropathy.<sup>14,15</sup>

Stent implantation in the systemic venous pathways has been performed in a limited number of patients with severe obstruction and symptoms.<sup>9-11</sup> In some, complete occlusion of the superior caval pathway required prior needle or stiff guidewire puncture for the initial recanalisation.<sup>18,19</sup> Both balloon expandable (Palmaz) and self expanding stents (Wallstents) have been used.<sup>9-11</sup> The self expanding stents,

which exert a lower force, seem more suited to obstruction caused by distortion and kinking of the pathway rather than scar formation or build up of intimal proliferation. When using self expanding stents, it is prudent to predilate the stenosis with a balloon to confirm that it is dilatable. On the other hand, when using balloon expandable stents, it is possible to use high pressure balloons and prior dilatation is not essential. Acceptable results have been achieved with resolution of the angiographic stenosis, a reduction in systemic venous pressure, and improvement in the clinical condition of patients. Follow up has been short but there are no reports of build up of significant amounts of neointima. In the pulmonary arteries, balloon expandable stents have been shown to be redilatable many months after implantation, so that modest amounts of neointimal proliferation in systemic venous stents may be amenable to repeat dilatation.<sup>20</sup> This may not apply to self expanding stents that cannot be overexpanded beyond their stated maximum diameter nor after adherence to the vessel wall.<sup>21</sup>

There are no randomised comparisons of stent implantation with balloon dilatation, and, given the numbers of patients available, this is unlikely to be possible in a single centre study. The effectiveness of stent implantation in the short term is also likely to discourage participation in such a study. The papers from Bu'Lock *et al* and Brown *et al* in this issue confirm, in two relatively large groups of patients, that the technique of stenting systemic venous pathway obstructions is safe.<sup>22,23</sup> Both balloon expandable and self expanding stents can produce a substantial increase in the size of the narrowed pathways. Malposition of stents is infrequent and does not seem to cause morbidity.

The indications for intervention, however, are considerably more liberal than would have been applied in the surgical era, and repeat surgery is unlikely to have been offered to many of these patients. Many of these patients were asymptomatic, with minimal gradients and modest stenoses (33–40%), and it is the availability of a safe and effective technique that has prompted intervention. Both papers justify the liberal indications by referring to a paper that they suggest indicates a significant correlation between sudden death and venous stenoses in asymptomatic patients. Review of this paper and others highlights that sudden death is unpredictable, and in some patients it only occurs in association with ventricular dysfunction or arrhythmia.<sup>2,3,6</sup> There are no studies directly linking venous pathway occlusion with sudden death. Indeed, in Bu'Lock *et al*'s series, five patients were given angiotensin converting enzyme inhibitors after the procedure—two have died and two are awaiting transplantation. Thus, liberally stenting the systemic venous pathways, while angiographically impressive, is unlikely to be beneficial with respect to these complications. Both studies report "subjective improvement" in exercise tolerance in some patients. There was no formal evaluation of functional ability before and after stent implantation; this information might support intervention in "asymptomatic" patients.

The need for transvenous pacemaker lead implantation in patients who are not symptomatic from venous obstruction is another reason for striving to preserve widely patent

pathways. The progressive loss of sinus rhythm over time suggests that an increasing number of patients will require pacemakers for bradycardia support or antitachycardia pacing. Indeed, the need for a transvenous electrode should prompt evaluation of the pathways so that moderate stenoses are not worsened by placement of transvenous electrodes.<sup>6</sup> Lead implantation appears to be facilitated by prior stent implantation.<sup>10 22 23</sup>

In the absence of evidence-based guidelines for stent implantation in asymptomatic patients with mild to moderate stenoses, it is mandatory that such patients are observed carefully for unexpected complications. Exercise tolerance should be documented before and after stent implantation. Inclusion into multicentre stent registries may allow a better evaluation of benefits and side effects.

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