Journal review

1. **Long term outcome of patients with isolated thin discrete subaortic stenosis treated by balloon dilation: a 25 year study.** De Lezo JS, Romero M, Segura J, et al. *Circulation* 2011;124:1461–8. doi: 10.1016/S0019-4832(12)60025-X

This 25 year follow-up, retrospective study describes results of percutaneous transluminal balloon tearing of isolated thin discrete subaortic stenosis (DSS) in 76 patients. The age of patients at the time of dilation ranged from 2–67 years (mean 19 ± 16 years). The inclusion for the study was restricted to only those patients who had isolated DSS where the subaortic membrane was thin (<3 mm thickness, without fibromuscular component) along with significant subaortic gradient or symptoms or ECG signs of left ventricular strain. Following balloon dilation, the left ventricular gradient decreased from 70 ± 27 mmHg to 18 ± 12 mmHg, ($P < 0.001$). No significant post procedure aortic regurgitation was observed. After a mean follow-up time of 16 ± 6 years, 11 patients (15%) developed restenosis, 3 patients (4%) progressed to muscular obstructive disease and one patient (1.3%) developed a new distant obstructive membrane. Twelve (16%) patients were redilated at a mean of 5 ± 3 years and 4 patients (5%) underwent surgery at a mean of 3 ± 2 years after their first treatment. Fifty-eight (77%) patients remained alive and free of redilation or surgery at follow-up. Larger annulus diameter and thinner membranes were independent factors associated with better long-term results. Authors concluded that most patients (77%) with isolated thin DSS treated with transluminal balloon tearing of the membrane had sustained relief at subsequent follow-up without restenosis, need for surgery, progression to muscular obstructive disease, or an increase in the degree of aortic regurgitation.

Perspective

Discrete subaortic stenosis is a rare cardiac lesion which was considered to be a mystery due to many uncertainties regarding its origin, pathogenesis, natural history, treatment options, and recurrence after treatment. Although DSS has classically been considered a congenital malformation, there is evidence for an acquired subaortic membrane during life. Though first described in 1985, balloon dilatation has not been widely used, perhaps due to variable results in literature. The thickness of the membrane and its proximity to the aortic valve and/or mitral valve are some of the factors which may influence the result of balloon tearing. The findings of this study in which patients have been followed up for a long period of time, may help in better selection of patients who can benefit from an interventional procedure as a first line management strategy. Though the recurrence rate is noticeable, it is similar to that observed in surgical patients and both are probably influenced by the progressive nature of the disease. If restenosis of the membrane develops, balloon dilation can be repeated successfully in most patients. With this strategy, surgery for a thin membrane may be delayed or even avoided.

2. **A comparison of Blalock-Taussig shunts with and without closure of the ductus arteriosus in neonates with pulmonary atresia.** Zahorec M, Hrubsova Z, Skrak P, Poruban R, Nosal M, Kovacikova L. *Ann Thorac Surg* 2011;92:653–9. doi: 10.1016/S0019-4832(12)60026-1

There is no consensus whether the patent ductus arteriosus (PDA) should be ligated or not at the time of performing modified Blalock-Taussig (BT) shunt in neonates with pulmonary atresia. This retrospective observational study from National Institute of Cardiovascular Diseases, Bratislava, Slovakia, aimed to address this issue. Between January 1997 and October 2010, 62 neonates (mean age 6.9 ± 5.5 days) underwent modified BT shunt through mid sternotomy approach. Of these patients 31 neonates underwent closure of PDA while in remaining half it was left open. The decision regarding closing or leaving PDA during surgery was based on institutional protocol in majority and on operating surgeon's preference, with nine different operators involved. Primary outcomes analysed were mortality, resuscitation events, and the need for reintervention within the first 48 postoperative hours.

Time to extubation, maximum vasoactive-inotropic score, and length of hospital stay were selected as the secondary outcome variables.

Compared with patients in whom the PDA was left open, patients with a surgically closed arterial duct had a higher incidence of resuscitation events (29.0% vs 0%, $P=0.0012$), reinterventions (35.5% vs 3.2%, $P=0.0013$), and higher early hospital mortality (9.7% vs 0%, $P=0.038$). Time to extubation and length of hospital stay did not differ between the 2 groups ($P=0.16$ and 0.73 , respectively). A trend towards a higher maximum vasoactive-inotropic score in the group with a closed duct was observed (median 13.5 vs 10, $P=0.10$). Authors concluded that in newborns with pulmonary atresia, ductal closure during modified BT shunt procedure is associated with increased incidence of resuscitation events, need for reintervention, and increased mortality during the early postoperative period.

Perspective

Systemic to pulmonary shunts have proven to be highly effective for the palliation of neonates with cyanotic congenital heart disease who are not candidates for early complete repair. Despite improvement in surgical techniques, operative mortality of modified BT shunt among neonates remains at 3–14%.

Though ductal patency may be lifesaving in case of early shunt obstruction or postoperative increase of pulmonary vascular resistance, it may also adversely influence the early postoperative haemodynamics due to increased diastolic run-off, volume overload, and low cardiac output state. Furthermore, this poses the risk of accompanying coronary steal and consequent impairment of myocardial contractility. According to the authors this could be avoided in the index study by choosing a smaller (3–3.5 mm) shunt. In contrast, in patients with closed PDA, episodes of hypoxaemia with or without documented shunt thrombosis are well known and were also observed by authors, leading to higher reintervention rate in this group of neonates. Though most surgeons in India refrain from closing PDA at the time of shunt surgery in neonates, this is the first study addressing the issue systematically. Although retrospective, the study provides data to support the approach of not closing PDA at the time of BT shunt in neonates.

3. Efficacy and safety of Bosentan for pulmonary arterial hypertension in adults with congenital heart disease.

Monfredi O, Griffiths L, Clarke B, Mahadevan VS. *Am J Cardiol* 2011;108:1483–8.

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The dual endothelin receptor antagonist, Bosentan, has been shown to be well tolerated and effective in improving pulmonary arterial hypertension (PAH) symptoms in patients with Eisenmenger syndrome but data from longer-term studies are lacking. The aim of this retrospective study was to analyse the long-term efficacy and safety of Bosentan in adults with PAH secondary to congenital heart disease (PAH-CHD). Prospectively collected data between October 2007 and June 2010 from adult patients with PAH-CHD (with and without Down's syndrome) initiated on Bosentan has been analysed. Parameters measured before Bosentan initiation (62.5 mg 2 times/day for 4 weeks titrated to 125 mg 2 times/day) and at each follow-up (1 month and 3, 6, 9, 12, 18, and 24 months) included exercise capacity (6-minutes walk distance [6MWD]), pre-test oxygen saturation, liver enzymes, and haemoglobin. Thirty-nine patients with PAH-CHD (10 with Down's syndrome) who had received >1 dose of bosentan (mean duration of therapy 2.1 ± 1.5 years) were included in the analysis. A significant ($P<0.0001$) average improvement in 6MWD of 54 m over a 2-year period in patients with PAH-CHD without Down's syndrome was observed. Male patients had a 6MWD of 33 m greater than females ($P<0.01$). This improvement in 6MWD was not observed in patients with Down's syndrome. In all patients, oxygen saturation, liver enzymes, and haemoglobin levels remained stable. None of the patients discontinued Bosentan due to adverse events. Authors concluded that patients with PAH-CHD without Down's syndrome gain long-term symptomatic benefits in exercise capacity after Bosentan treatment. Men seem to benefit more than women. Bosentan appears to be well tolerated in patients with PAH-CHD with or without Down's syndrome.

Perspective

Bosentan is the only endothelin receptor antagonist currently recommended for patients with Eisenmenger syndrome. However, there is limited data regarding sustained beneficial effects of bosentan in these patients. This study from University of Manchester, United Kingdom addresses this issue in adults with PAH-CHD. Sustained benefits and that too without significant side effects seen in the study are promising considering otherwise unfavourable natural history of CHD-PAH. However, it must be noted that the average age of patients was 38.7 ± 13.3 years. The functional class at baseline or on follow-up is not mentioned in the study. Though difficult to hypothesise the underlying mechanism, males did better than females in terms of 6MWD improvement. In concordance with other studies, there was no change in oxygen saturation. Data from this study and the earlier BREATH 5 trial prompts increasing use of bosentan in all patients with Eisenmenger syndrome. However, current European Society of Cardiology (ESC) guidelines limit use of bosentan in patients with World Health Organization (WHO) functional class III or more. These guidelines assume even more significance in Indian context since bosentan is a rather expensive drug.