

## Heartbeat: New insights in congenital heart disease

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Overall survival of patients with transposition of the great arteries (TGA) now is about 97% at 25 years because of the success of the arterial switch procedure, starting in the 1980's. However, patients require ongoing monitoring, medical therapy and, in some cases, reoperation due to late sequelae such as aortic sinus dilation, aortic valve dysfunction, right ventricular outflow obstruction or arrhythmias.<sup>1</sup> Pulmonary arterial hypertension (PAH) is a rare, but highly morbid, associated condition in TGA patients after an arterial switch procedure. In a series of 25 children with PAH after a TGA arterial switch repair, only 58% survived 5 years or longer despite PAH-targeted medical therapy in 19 patients, lung transplantation in four patients and the addition of a Potts shunt in two patients.<sup>2</sup> Prognosis was equally poor in those with early onset PAH (within weeks to months) and those with late-onset PAH. (figure 1)

In the accompanying editorial, Lammers and Bauer present data from the German National Register for Congenital Heart Defects database showing a prevalence of PAH in TGA patients with an arterial switch repair of only 0.11% (95% CI 0.29% to 0.64%).<sup>1</sup> Even though rare, they suggest that this association “suggests that alternative mechanisms for the development of PAH may exist in congenital heart disease, thus complementing the established pathogenic concept focusing entirely on long-standing increase in pulmonary blood flow leading to pulmonary vascular disease and pulmonary hypertension.”

Body mass index (BMI) has been associated with cardiovascular mortality for several types of heart disease but has not previously been studied in adults with congenital heart disease (ACHD). In a cohort of over 3000 ACHD patients, a high BMI was associated with a slightly better prognosis, after adjustment for age, defect complexity, cyanosis and objective measures of exercise capacity.<sup>3</sup> Weight loss also was associated with higher mortality. Although it makes sense that underweight patients have lower survival rates, likely related to cardiac cachexia, it seems counterintuitive that overweight-obese patients had higher

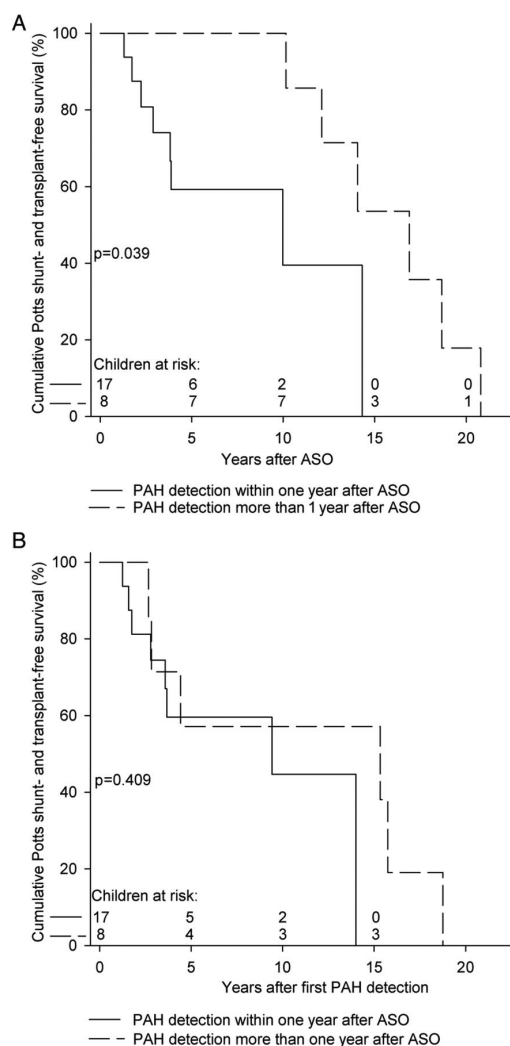
**Figure 1** Potts shunt- and transplant-free survival of children with first PAH detection within and more than 1 year after ASO. (A) Survival from ASO. Survival of 1, 3, 5 and 10 years was 100%, 74%, 59% and 40% for the 18 children with first PAH detection within 1 year after ASO and 100%, 100%, 100% and 100% for the 7 children with first PAH detection more than 1 year after ASO, respectively ( $p=0.039$ ). (B) Survival from first PAH detection. Survival of 1, 3, 5 and 10 years was 100%, 75%, 60% and 45% for the 18 children with first PAH detection within 1 year after ASO and 100%, 71%, 57% and 57% for the 7 children with first PAH detection more than 1 year after ASO, respectively ( $p=0.409$ ). ASO, arterial switch operation; PAH, pulmonary arterial hypertension.

survival rates compared with those with a normal BMI. (figure 2) However, additional analysis revealed a U-shaped curve with higher mortality in underweight and severely overweight patients, compared with the median BMI of 24 kg/m<sup>2</sup>. (figure 3)

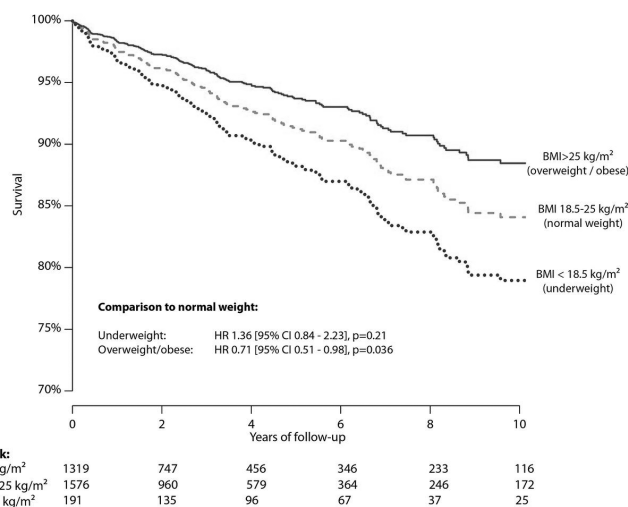
In a commentary, Burden focuses on the association between weight loss and higher mortality in ACHD patient and raises the question whether cachexia should be actively managed in these patients.<sup>4</sup> He also suggests “clinical trial evidence is needed on the safety

of weight reduction diets in the obese and overweight person with ACHD. Meanwhile, attention to exercise is more appropriate.”

In men, erectile dysfunction (ED) is associated with an increased risk of cardiovascular disease however there is little data on the association between treatment for ED after myocardial infarction (MI) and cardiovascular outcomes. In nationwide Swedish cohort of over 43 thousand men hospitalised for first MI, the 7.1% who received ED medications over the subsequent 3



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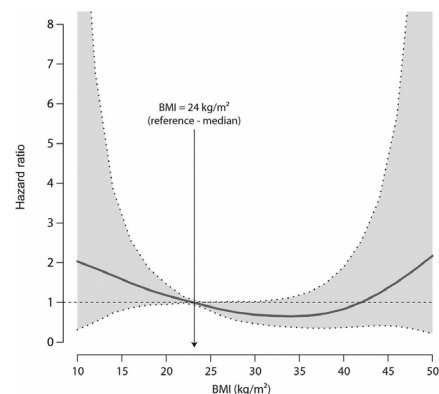
**Figure 2** Survival by body mass index (BMI, kg/m<sup>2</sup>) based on the results of the Cox analysis. Compared with normal weight patients (ie, BMI 18.5–25 kg/m<sup>2</sup>), underweight patients tended to have worse outcome, while overweight/obese patients as a group had on average a significantly superior prognosis.

years had a 33% lower mortality and 40% lower risk of heart failure hospitalisation compared with those who did not received ED medications.<sup>5</sup> This effect appeared to be dose-dependent and was only seen with phosphodiesterase-5 (PDE5) inhibitors. (figure 4)

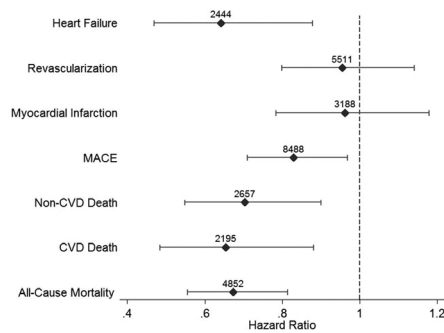
Prof. Collinson<sup>6</sup> discusses the limitations of this study and the potential safety concerns in patients with heart disease taking nitrates. He summarises his comments as follows: “The first conclusion

is that treatment with PDE5 inhibitors appears to be safe. Therefore, cardiologists may recommend PDE5 inhibitors to their patients (with appropriate clinical assessment and medication review). The second is much more intriguing. PDE5 inhibitors may be cardioprotective and a new therapeutic option for treatment or prevention.”

Review articles worth reading in this issue of *Heart* include *Palliative care in patients with end-stage valvular heart*



**Figure 3** Association between HR of death and BMI as a continuous variable, illustrating excess mortality in underweight and severely obese patients. The median BMI of the population (24 kg/m<sup>2</sup>) was set as the reference value.



**Figure 4** Adjusted HRs and 95% CI for the association between treatment for erectile dysfunction, compared with no treatment for erectile dysfunction, and outcomes after a first myocardial infarction in 43 145 men. Number of events are depicted above the point estimate for each outcome. MACE, major adverse cardiac event; CVD, cardiovascular disease.

*disease*<sup>7</sup> and *Value in Cardiovascular Care*.<sup>8</sup> These articles will expand your knowledge in areas core to the practice of clinical cardiology that we often fail to discuss enough among ourselves.

For the *Cardiology in Focus* feature in this issue, I have written a short summary<sup>9</sup> entitled ‘Tips for publishing your clinical cardiology research’. A detailed table summarises the essential elements and ‘tips’ for preparing each section of your research paper from title to discussion. We hope that future authors will use this table as a guide to improving the likelihood that your research will be accepted for publication in *Heart*.

**Competing interests** None declared.

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