Heartbeat: Highlights from this issue

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As increasing numbers of patients with congenital heart disease (CHD) survive the neonatal period and reach adulthood we are faced with a growing patient population, both, in terms of numbers and complexity of disease. The vast majority of patients are not cured and requires lifelong specialized medical care. The current focus issue on congenital heart disease combines publications dealing specifically with this challenging group of patients. It highlights the heterogeneity of problems faced by congenital cardiologists, spanning from a changing diagnostic spectrum to issues of risk stratification, haemodynamic assessment as well as the growing issue of pregnancy in patients with congenital heart disease.

The Editor's choice in this issue is a prospective study by Balci and colleagues (***see page 1373***). The authors aimed to prospectively validate three published risk models of adverse outcome during pregnancy in women with congenital heart disease. Based on a sample of 213 pregnancies in 203 women, the authors investigated prospectively, the validity of three commonly used risk scores for maternal and offspring risk. The scoring instruments included were the ZAHARA I, the CARPREG score and the WHO classification of maternal cardiovascular risk. The authors show that all three scoring systems have important limitations when predicting risk to mother and child. This is illustrated by the results of the area under curve analysis highlighting the only modest discriminatory ability of the scoring systems (Figure 1).

In the accompanying editorial Drs Diller and Uebing (***see page 1311***) remind us about the limitations of risk scores in the setting of congenital heart disease in general, and the fact that "all available risk scores can only roughly guide the clinician when counseling women with [congenital heart disease] of childbearing age". This is not only due to the fact that most scores are not externally validated and adjusted to specific settings, but is also related to the problem that some important risk factors are not included in the scoring systems. Therefore, patients with congenital heart disease benefit from expert assessment including the individual estimation of risk based on anatomy, previous therapeutic efforts and history of complications.

Risk stratification in patients with pulmonary hypertension, both idiopathic and as a consequence of congenital heart disease or Eisenmenger syndrome also remains challenging. Increasingly, tools for risk stratification and for guiding medical therapy are required. Two papers address this important and timely issue. While Dr Scognamiglio and colleagues (***see page 1335***) show how a widely available laboratory marker of inflammation – C-reactive protein (CRP) – represents powerful a marker of mortality in this setting and could be incorporated in the routine assessment of these patients, Di Maria *et al* (***see page 1342***) propose a novel indicator of right ventricular performance, integrating contractility, afterload and right ventricular-to-vascular coupling as a potential risk predictor. The authors assess the utility of this novel parameter in 50 children with pulmonary hypertension and report an association with abnormal WHO class, the need for atrial septostomy, as well as mortality. This article nicely illustrates the dynamic nature and complex physiology of pulmonary hypertension. The editorial by Lammers *et al*. (***see page 1305***) reminds us that "no risk marker should be seen in isolation and that the degree of pulmonary hypertension (i.e. the absolute value of mean or systolic pulmonary arterial [PA] pressure) per se is insufficient to predict the outcome of patients" as appreciated by experienced clinicians.

If patients with congenital heart disease and complex lesions are left untreated, some will progress to Eisenmenger syndrome, a condition with poor prognosis and still limited treatment options as highlighted by a systematic review of the literature and retrospective analysis of data from a large European single centre as well as a review article in the current issue (***see pages 1366, 1373***). The problem of increasing numbers of patients with complex congenital heart disease – such as those with univentricular physiology – surviving due to early surgical interventions is also the topic of a study by Coats *et al* (***see page 1348***). The authors point out that,

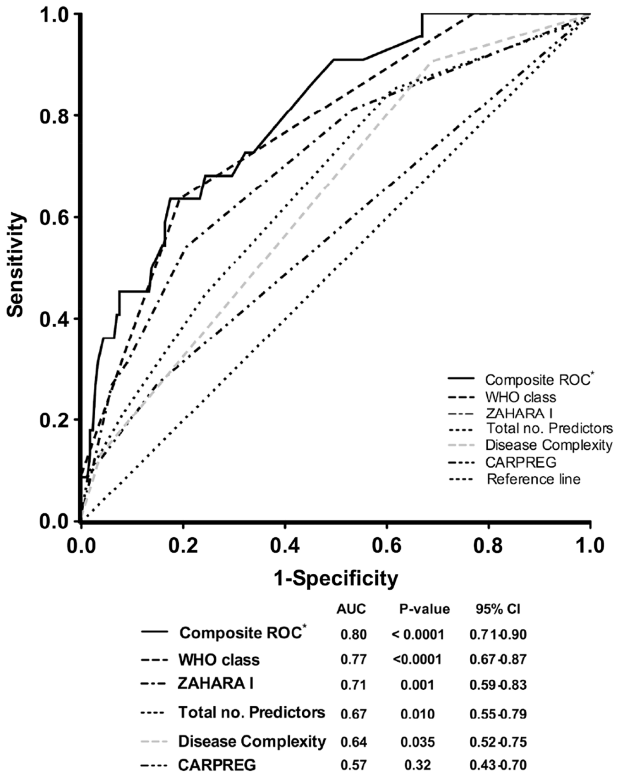

Figure 1 Receiver Operating Characteristic Curves of maternal cardiovascular events for the different cardiovascular risk assessment models. (AUC, area under the curve; CI, confidence interval; ROC, receiver operating characteristic; WHO, World Health Organization).

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according to their estimates, the current population of patients with univentricular physiology in the UK consists of over 1000 adults and approx. 1700 children. More importantly, the authors estimate that "adult numbers will increase by over 60% in the next decade with the proportion in NHYA functional class 3 set to double". This will of course pose huge logistic challenges regarding the workforce of trained (paediatric and adult) congenital heart disease specialists required to care for this patient population. In the accompanying editorial Dr Bhatt also draws our attention to the fact that state of the art care requires adequate resources, quality improvement and future investments in education (***see page 1308***).

Other clinically challenging topics are also addressed in this CHD focus issue. This includes the problem that an increasing number of patients after arterial switch operation for transposition of the great arteries (TGA) present with aortic dilatation. It is currently unknown whether aortic dilation in this setting should prompt proactive aortic root replacement. Van der Bom and colleagues (***see page 1360***) suggest that neo-aortic dilatation is a common problem in TGA patients and aortic growth does not seem to stabilize over time in early adulthood. These findings provide a rationale for periodic imaging of aortic dimensions in these young patients but are – in isolation – insufficient to warrant early surgical repair. Clinicians should also not confound statistically significant dilation with a clinically meaningful one. Therefore, these novel data are important but require further confirmation and especially correlation with clinical events before they should prompt a change in clinical practice.

In addition, Radke and Baumgartner (***see page 1382***) provide a comprehensive overview over the diagnosis and treatment of Marfan syndrome in the Education in Heart section. This is a challenging area that has received increasing interest recently with emerging new therapeutic options such as the potential benefit of ACE-Inhibitor and ARB therapy. In the Image Challenge section Rommel *et al* (***see page 1392***) report about an unusual case of syncope in a young patient with absent pericardial sac.

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