

# Cardiology in the Young

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### **Development and validation of a conductance method to measure the impact of loading conditions on pulmonary regurgitation**

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Pulmonary regurgitation is a significant determinant of right ventricular and global functional performance late after Fallot's repair. We have previously quantified pulmonary regurgitation using pressure volume loops by measuring right ventricular volume increase during the isovolumic relaxation period. This (right ventricular angiogram digitisation) and other methods of measurement of right ventricular volume (e.g. magnetic resonance imaging) do not allow real-time analysis. However, right ventricular blood volume measurement by a conductance catheter may allow beat-to-beat analysis of right ventricular volume and pulmonary regurgitation fraction. Five silicone casts of human right ventricles of known volume were constructed and their volume was measured by the conductance catheter. The conductance catheter was found to be highly accurate and linear ( $r^2 0.94 \pm 0.02$  (SD); Y intercept  $-0.23 \pm 1.7$  ml). Thirteen patients (8 tetralogy of Fallot; 2 tetralogy of Fallot + atrioventricular septal defect; 3 open pulmonary valvotomy) late after surgery (3-35 years) had pulmonary regurgitation measured and indexed to stroke volume by (1) magnetic resonance imaging (spontaneous breathing) and (2) at right heart catheterisation (general anaesthesia) using simultaneous microtip pressure and conductance volume measurement. There was a linear relation between pulmonary regurgitation measured by magnetic resonance imaging and conductance catheter ( $r^2 0.59$ , Y-intercept 2.2%,  $p < 0.01$ ). During catheterisation, intrathoracic pressure was increased by a Valsalva manoeuvre (20 cm H<sub>2</sub>O); this increased pulmonary regurgitation ( $16.3 \pm 11.4\%$  (SD) to  $25.7 \pm 17.3\%$ ,  $p < 0.01$ ) and tended to decrease stroke volume ( $p = 0.06$ ). Transient right pulmonary artery occlusion was performed in 7 patients by inflating a 20 ml sizing balloon, this increased right ventricular end systolic pressure ( $69.1 \pm 21.4$  to  $78.7 \pm 23.1$  mm Hg,  $p < 0.05$ ) and increased pulmonary regurgitation ( $27.5 \pm 11.3$  to  $36.9 \pm 12.8\%$ ,  $p < 0.05$ ). In summary, conductance volume measurement allows real-time dynamic assessment of pulmonary regurgitation and demonstrates that pulmonary regurgitation is increased by positive intrathoracic pressure and branch pulmonary artery stenoses.

### **Anthracycline-induced changes in diastolic filling are related to differential effects on long- and short-axis left ventricular function**

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Left ventricular myocytes are arranged with the circumferential fibres sandwiched between two layers of longitudinal fibres, which may be differentially affected by pathological processes. The subendocardium is known to be particularly susceptible to myocardial insults, and anthracycline-induced myocardial damage is often predominantly subendocardial, particularly at low doses. Diastolic transmitral Doppler flow patterns and digitised M-mode traces of left ventricular long and short-axis wall motion were studied serially in seven children receiving Epirubicin for soft tissue sarcomas; prior to initial anthracycline exposure and every 100 mg/m<sup>2</sup> thereafter, to between 270 and 600 (median 500) mg/m<sup>2</sup>. Median fractional shortening fell throughout treatment, from 34 to 31.5% ( $p = 0.04$ ), with a similar reduction in circumferential fibre shortening rate and increase in end-systolic wall stress. There were no significant changes in peak rates of either systolic or diastolic short-axis wall motion or of longitudinal systolic shortening. In contrast, striking changes in both magnitude and peak rates of longitudinal diastolic lengthening were seen, particularly at low anthracycline doses. Although the absolute magnitude of early diastolic lengthening remained relatively constant, atrial phase lengthening increased with the first 100 mg/m<sup>2</sup>, and then fell markedly at higher doses, accompanied by a reduction in the peak rate of early diastolic lengthening and increase in the duration of early lengthening. Thus the total magnitude of diastolic lengthening also fell between 1 and 300 mg/m<sup>2</sup>. Atrial phase trans-mitral flow was increased after 100 mg/m<sup>2</sup>, and fell again at higher doses, along with a reduction in rate and time of deceleration from peak early filling velocity and heart rate. At low doses, anthracycline-induced myocardial damage is associated with Doppler echocardiographic findings consistent with altered myocardial relaxation, and with abnormal atrial phase long-axis lengthening. The simultaneous occurrence of such abnormalities suggests a common cause; perhaps initial differential damage of subendocardial myocytes. Such functional/pathological correlates may well have broader implications and merit further study.

### Pulmonary atresia with intact septum: growth of the right ventricle and tricuspid valve following primary procedure

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The initial surgical strategy for pulmonary atresia with intact ventricular septum is critical not only for survival but also for the growth potential of the right ventricle and tricuspid valve. As part of the UK and Eire Collaborative Study of Pulmonary Atresia with Intact Ventricular Septum (an on-going population based study of all children born in the UK and Eire since January 1991), we examined the growth of the tricuspid valve and right ventricle following primary procedure. Serial echocardiograms were reviewed in 61 children both pre- and post-primary procedure (mean 17 months post-procedure, range 2-45). The annulus of the tricuspid valve and length of the right ventricular inlet were measured in the four-chamber view. Z-scores were derived from published nomograms based on surface area. The initial tricuspid valve Z-score was significantly smaller in those undergoing shunt than in those undergoing various right ventricular outflow tract procedures with or without concomitant shunt ( $-2.2 \pm 0.3$ ,  $-0.8 \pm 0.2$  respectively,  $p=0.0002$ , data shown as mean  $\pm$  SEM). The change in tricuspid valve Z-score post-procedure, however, was similar (shunt  $-0.7 \pm 0.4$ , right ventricular outflow tract  $-0.8 \pm 0.2$ ,  $p=NS$ ). Similarly, the right ventricular inlet Z-score was smaller in the shunt group than the right ventricular outflow tract group ( $-1.6 \pm 0.1$ ,  $-1.2 \pm 0.1$  respectively,  $p=0.0009$ ). Again, the change in the Z-score was similar (shunt  $-0.06 \pm 0.1$ , right ventricular outflow tract  $-0.3 \pm 0.1$ ,  $p=NS$ ). Of those patients undergoing right ventricular outflow tract procedures alone, neither type of procedure (catheter versus surgery), nor requirement for a shunt within 6 weeks of procedure affected the change in tricuspid valve or right ventricular inlet Z-score. However, in patients with primary right ventricular outflow tract procedure, change in tricuspid valve and right ventricular inlet Z-score was weakly correlated with tricuspid valve and right ventricular Z-scores pre-procedure. Larger increases in tricuspid valve and right ventricular inlet size post-procedure were associated with smaller initial tricuspid valve and right ventricular inlet sizes ( $r=0.47$ ,  $p=0.01$  and  $r=0.44$ ,  $p=0.01$  respectively). There was no such correlation in patients shunted alone. Following establishment of right ventricular to pulmonary artery continuity, growth of the tricuspid valve and right ventricle frequently does not exceed somatic growth but is more likely in those patients with small initial tricuspid valve and right ventricle.

### Optimising oxygen delivery after paediatric cardiac surgery: should transfusion practice change?

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In the critically ill patient, optimising oxygen delivery can improve outcome. Oxygen delivery is influenced by both haemoglobin concentration and blood volume. Detection of anaemia and hypovolaemia in infants is difficult however, and laboratory and clinical assessment can be unreliable. The indications for transfusion ("triggers") and haematological and physiological aims of transfusion ("targets") are often poorly defined. We hypothesise that current transfusion practice fails to optimise oxygen delivery in children after cardiopulmonary bypass surgery. Prospective study of infants <1 year or <10 kg undergoing elective cardiopulmonary bypass. Daily measurements of red cell volume (biotin labelled red cell reinjection technique), haemoglobin haematocrit and haemodynamic variables were made once preoperatively and for 4 days postoperatively. Blood volume was derived from red cell volume and haematocrit values. Perioperative management followed standard departmental protocols. We also studied the first blood transfusion given once the patients were considered stable after surgery and the effect it had on red cell volume, blood volume, haemoglobin, haematocrit and haemodynamic variables. Measurements were made once prior to transfusion and repeated 24 hours later. Transfusion

"triggers" and "targets" were recorded. Twenty-six infants were studied: age 0.25-23 (median 5.25) months; weight 2.9-9.2 (median 5.7) kg; cardiopulmonary bypass time 32-174 (median 97) min. One child did not require a blood transfusion during the study period. Five children were transfused only in the initial postoperative period whilst continuing to bleed therefore had to be excluded. Primary "triggers" for blood transfusion included low haemoglobin, hypotension, suboptimal filling pressures, oliguria and acute blood loss. Pretransfusion red cell volume was low ( $<30$  ml/kg) in 16 of our 20 patients and blood volume was low ( $<85$  ml/kg) in 17/20. After transfusion, all had haemoglobin  $>13$  g/dl and although red cell volume measurement was unsuccessful in 2 children, values remained low in 11/18, range 12.8-44.5 (median 27.4) ml/kg. Blood volume was suboptimal in 16/18, range 28-106 (median 60) ml/kg. Hypovolaemia and low red cell volume are common after paediatric cardiopulmonary bypass. "Triggers" for transfusion are inconsistent and "targets" are ill defined. There is a tendency to undertransfuse resulting in suboptimal oxygen delivery. Prevention or correction of occult anaemia and hypovolaemia with a more generous transfusion regimen will improve oxygen delivery and may reduce morbidity and the need for intensive care.

### Pre- and postoperative determinants of restrictive right ventricular physiology after repair of tetralogy of Fallot

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We have recently reported that restrictive right ventricular physiology late after tetralogy of Fallot reduced pulmonary regurgitation and improved exercise capacity. Conversely, non-restrictive physiology led to right ventricular dilation and QRS-prolongation, a sensitive marker of late arrhythmias. The aim of this study was to assess determinants of restrictive right ventricular physiology in patients with tetralogy of Fallot undergoing repair during the last 10 years. We have studied 88 patients 1 month to 9 years (median 1.6 years) after repair with Doppler echocardiography. Age at surgical repair was from 2 months to 43 years (1.9 years). Restrictive right ventricular physiology defined as antegrade diastolic pulmonary artery flow with atrial systole was found in 28 patients (32%), and was unrelated to age at repair, previous shunt, crossclamp or bypass time. However, 16 of 39 patients (41%) with transannular patch had a restrictive right ventricle as compared to 2 of 25 (8%) with outflow tract repair only ( $p<0.05$ ). The QRS duration (ms) in patients with restrictive and nonrestrictive right ventricular physiology was related to the type of right ventricular outflow tract repair:

	Transannular patch	Monocusp	Outflow patch
Restrictive	119.8 $\pm$ 18.6	116.3 $\pm$ 18.9	124.0 $\pm$ 6.9
Non-restrictive	133.3 $\pm$ 12.2	121.4 $\pm$ 32.0	123.1 $\pm$ 18.4
	$p<0.02$	NS	NS

Restrictive right ventricular physiology is more common in patients requiring a transannular patch. Nonetheless, nonrestrictive right ventricular physiology in the presence of a transannular patch significantly increases QRS duration with its potential deleterious long-term effects.

### Negative pressure ventilation using the Hayek external high frequency oscillator cuirass improves cardiac output after right heart surgery

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The low cardiac output state in children after cardiopulmonary bypass often occurs as a consequence of reduced pulmonary blood flow. This is best demonstrated in postoperative patients after surgery to the right heart. Although often a transient problem it represents a difficult management challenge. In this study we have investigated the potential use of negative pressure ventilation as a haemodynamic tool in children after right heart surgery. We studied the effect of negative pressure ventilation on cardiac output in a group of ten paralysed and fully sedated children (median age 5.3 years) in the early postoperative period

following fenestrated total cavopulmonary connection (n=5) or total correction of tetralogy of Fallot (n=5). All patients were initially ventilated with volume-cycled intermittent positive pressure ventilation. Negative pressure ventilation was delivered using the Hayek external high frequency oscillator cuirass. Cardiac output was calculated using the direct Fick method. Oxygen consumption was measured by bedside mass spectrometry using the inert gas dilution method. All children had indwelling pulmonary arterial lines for measurement of mixed venous oxygen content. Arterial oxygen content was calculated from peripheral arterial, left atrial or pulmonary venous blood samples where appropriate. Cardiac output was measured in a cardiorespiratory state on intermittent positive pressure ventilation, and after 15 minutes of negative pressure ventilation. The rate and inspiratory pressure during negative pressure ventilation were adjusted to maintain similar end-tidal carbon dioxide readings, and the inspired oxygen concentration was not changed. Negative pressure ventilation increased the cardiac output by a mean of  $47 \pm 17$  (SD)% ( $p < 0.02$ ). In the children with the Fontan circulation the mean increase was  $43 \pm 10.3\%$ , and in the group with tetralogy of Fallot was  $49 \pm 21.6\%$ . Mixed venous saturation increased by a mean of  $6.4 \pm 4.8\%$  ( $p = 0.01$ ), as did mixed venous oxygen content ( $p < 0.02$ ). There was no change in the mean arterial pressure, pulmonary arterial pressure or heart rate. Negative pressure ventilation leads to a highly significant increase in cardiac output after right heart surgery and may prove to be an important therapeutic option in children with low-output states.

#### Clinical assessment of ductal shunting in the preterm: the significance of the murmur

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It has been known for many years that a large left-to-right ductal shunt may be present in preterm infants without a heart murmur, yet most referrals for cardiological assessment in preterm babies arise because a murmur has been detected. We aimed to find out why the murmur usually arises, and when present if it is associated with a larger left-to-right shunt than in those without. 442 echocardiograms were reviewed from 127 preterms ventilated for respiratory failure. 117 scans showed clear evidence of a large left-to-right shunt, with a widely patent duct and left atrial:aortic root ratio  $> 1.4:1$ . Fifty-five scans associated with a murmur were compared with 62 without. The pattern of ductal flow through the cardiac cycle and left ventricular output (which increases with left-to-right shunting) were compared between the two groups. Ductal flow patterns were categorised where possible into bidirectional (group A), or pure left-to-right shunt with low velocity in systole and high in diastole (B), pure left-to-right shunt with high velocity in systole and low in diastole (C), and pure left-to-right shunt with continuous high velocity (D). Results: *Pattern of flow within the duct.* Pattern type C (high in systole, low in diastole) occurred in two thirds (66%) of babies with a murmur but only in 18% of those without ( $p < 0.01$ ), amongst whom the commonest pattern was bidirectional (type A, 36%). *Left*

*ventricular output.* Amongst babies with a murmur, mean (SD) left ventricular output was 501 (108) mls/kg/min; much higher than 352 (99) mls/kg/min amongst those without a murmur ( $p < 0.01$ ). The difference was related to stroke volume more than heart rate. This study suggests that the typical murmur associated with left-to-right ductal shunting in the preterm usually develops when high volume flow through the duct is restricted to systole. "Silent" ductal shunting as detected by a dilated left atrium on echocardiography is usually associated with a smaller left-to-right shunt than in babies with an associated heart murmur and otherwise similar M-mode and cross-sectional echocardiographic features.

#### Transcatheter occlusion of the patent arterial duct with Cook detachable coils

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Transcatheter occlusion of the patent arterial duct in children and adults is a common alternative to surgery. Concerns exist about residual shunts, up to 38% at one year with the Rashkind double umbrella technique. Gianturco spring coils positioned from the aorta or antegradely via the pulmonary artery are an increasingly used option. Embolisation to branch pulmonary arteries has occurred in up to 9%. We report our experience with a new Cook detachable coil system for occlusion of patent arterial ducts. Sixty-six consecutive patients, 21 male, 45 female, aged 1.2-22 years, with a patent arterial duct, underwent elective transcatheter closure. Forty-two of these were native patent arterial ducts (Group A) with a minimum diameter of 1.0-5.0 mm (median 2.2mm). A further 24 had undergone previous occlusion procedure (Group B). A transvenous approach was used in the majority of patients with a 4-6.3 Fr. delivery catheter. A transarterial route was solely used in 12 patients. A total of 127 detachable Cook coils were successfully implanted in 65 patients with a median of 2 coils in Group A and 1 coil in Group B. Complete occlusion of native patent arterial ducts, assessed by colour flow echocardiography, was achieved in 37/42 (88%) at 24 hours, 38/42 (91%) at 1 month and 40/42 (95%) by 6 months follow-up. One (1.5%) embolisation, of a 5mm coil, occurred in a child with a 2.5mm diameter duct. Eight coils were electively removed due to poor positioning. One child has undergone a further coil procedure 9 months later for a residual leak resulting in complete closure. Another has a residual leak at one month post implantation. In one patient in Group B the lesion could not be crossed by a 4F catheter and the procedure was abandoned. Of the remaining 23, 87% were occluded at 24 hours and 21/23 (91%) at one month post implantation. An increase in post implantation left pulmonary artery velocities was seen in two patients. Transcatheter occlusion using detachable Cook coils is a safe and effective alternative to presently available devices. The delivery system benefits from being fully retrievable until a satisfactory position is obtained.