Pulmonary and caval flow dynamics after total cavopulmonary connection

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Abstract

Objective—**To assess flow dynamics after total cavopulmonary connection (TCPC).**
Design—**Cross-sectional study.**
Setting—**Aarhus University Hospital.**
Patients—**Seven patients (mean age 9 (4–18) years) who had previously undergone a lateral tunnel TCPC mean 2 (0.3–5) years earlier.**
Interventions—**Pressure recordings (cardiac catheterisation), flow volume, and temporal changes of flow in the lateral tunnel, superior vena cava, and right and left pulmonary arteries (magnetic resonance velocity mapping).**
Results—**Superior vena cava flow was similar to lateral tunnel flow (1.7 (0.6–1.9) *v* 1.3 (0.9–2.4) l/min★m^2) (NS), and right pulmonary artery flow was higher than left pulmonary artery flow (1.7 (0.6–4.3) *v* 1.1 (0.8–2.5) l/min★m^2, p < 0.05). The flow pulsatility index was highest in the lateral tunnel (2.0 (1.1–8.5)), lowest in the superior vena cava (0.8 (0.5–2.4)), and intermediate in the left and right pulmonary arteries (1.6 (0.9–2.0) and 1.2 (0.4–1.9), respectively). Flow and pressure waveforms were biphasic with maxima in atrial systole and late ventricular systole.**
Conclusions—**Following a standard lateral tunnel TCPC, flow returning via the superior vena cava is not lower than flow returning via the inferior vena cava as otherwise seen in healthy subjects; flow distribution to the pulmonary arteries is optimal; and some pulsatility is preserved primarily in the lateral tunnel and the corresponding pulmonary artery. This study provides in vivo data for future in vitro and computer model studies.**
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Total cavopulmonary connection (TCPC) is widely used as final palliation of many complex congenital heart malformations.[1] It is dependent on low pressure loss through the cavopulmonary circulation as well as on optimal flow distribution to the lungs.[1 2] In vitro studies and computer simulations aiming to define the optimal geometry of the TCPC circulation have been performed and results implemented clinically.[2–6] Simulations depend on input parameters such as pulsation characteristics, absolute flow, and flow ratios in the involved vessels, but in vivo information on these parameters is sparse.[2 7]

Flow in the TCPC has usually been considered passive and non-pulsatile.[8 9] All simulations have therefore been based on steady inflow parameters and the assumption that flow returning from the inferior vena cava is much higher than flow returning through the superior vena cava.[2–6] Lack of pulsation may theoretically be a drawback as experimental and clinical studies indicate that steady, non-pulsatile pulmonary flow may increase vascular resistance,[10–12] promote development of pulmonary arteriovenous fistulae,[10 12 13] and attenuate pulmonary arterial growth.[12 15 16] Although it is generally accepted that right atrial contraction is not essential after a Fontan-type operation, it may, nevertheless, play an important role by generating pulsatile flow.[8 17] It is, however, possible that the intra-atrial position of a lateral tunnel, as well as contraction of the part of the lateral tunnel consisting of atrial musculature, may induce some pulsatility in the TCPC.

Using magnetic resonance (MR) velocity mapping as an accurate, non-invasive method for measurement of volumetric flow,[18–21] cardiac catheterisation, and angiography, we performed a study of the in vivo blood flow characteristics of the TCPC circulation.

Methods

PATIENTS

Seven patients (four male, three female) (mean age 9 (4–18) years), who underwent TCPC for tricuspid (n = 5) or mitral (n = 2) atresia, mean 2 (0.3–5) years earlier, were studied. The TCPC (fig 1) included an anastomosis between the superior vena cava and the right pulmonary artery in six patients, and between a single left superior vena cava and the left pulmonary artery in one patient (patient 7). Via an intra-atrial lateral tunnel made of Gore-Tex, which included part of the right atrial wall, the inferior vena cava was connected either directly to the right pulmonary artery (n = 2) or to the transected main pulmonary artery (n = 5) (table 1). The horizontal offset between the two cavopulmonary anastomoses was approximately one vessel diameter with the superior vena cava to the right, except in two subjects, one (patient 2) with no offsetting and one (patient 7) with a left superior vena cava. Three (patients 1, 2, and 7) had fenestrations in the patch.

Five patients (patients 1, 2, 3, 4, and 5) were in New York Heart Association (NYHA) class I–II, one (patient 6) was in class III, and one
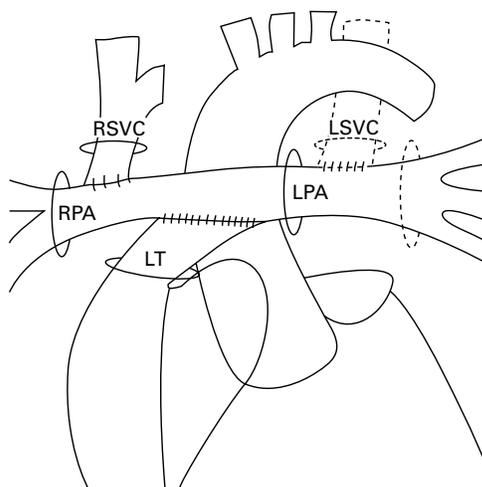
Figure 1 Schematic drawing of the TCPC and the planes used for MR flow measurements. LPA, left pulmonary artery; LSVC, left superior vena cava; LT, lateral tunnel; RPA, right pulmonary artery; RSVC, right superior vena cava

MR scanning

MR scanning was performed on a Philips 1.5 Tesla Gyroscan S15/HP whole body scanner (Philips Medical Systems, Best, Netherlands). Patients were examined in the supine position after an initial series of spin echo scouts had been obtained to localise the vessels under study.

MR velocity measurements were performed using the flow adjusted gradients sequence,[22] through the following four imaging planes (fig 1): (1) lateral tunnel—at the cranial aspect of the right atrium; (2) superior vena cava—one vessel diameter above the pulmonary artery anastomosis; (3) right pulmonary artery—one vessel diameter lateral to the superior vena cava anastomosis; and (4) left pulmonary artery—one vessel diameter lateral to the pulmonary bifurcation. In the patient with a left superior vena cava (patient 7), the left pulmonary artery measurement was performed one vessel diameter lateral to the superior vena cava anastomosis. For each flow measurement, the field of view was 350 mm and 128 phase encoding steps were obtained, yielding a pixel size of 2.7×2.7 mm. Slice thickness was 8–10 mm. Other imaging parameters were two signal averages, echo time 9–12 ms, flip angle 45°, and maximum velocity 1 m/s. Eighteen to 27 time frames were obtained with a repetition time of 25 to 28 ms covering 80–100% of the cardiac cycle. Electrocardiographic triggering was used. The total number of heart beats required to obtain data for one flow measurement was 512. Respiratory gating was omitted to set off the haemodynamic effects of respiration. The total examination time was approximately 60 mins depending on heart rate.

DATA ANALYSIS

Cardiac catheterisation

From the angiograms, the direction of preferential streaming from the superior vena cava and the lateral tunnel was visually determined by one observer blinded to the MR data. The pulmonary artery primarily receiving the superior vena cava or lateral tunnel flow was defined as the corresponding pulmonary artery (PAc). From the pressure tracings, R–R intervals were divided into 20–25 intervals and instantaneous pressure differences between the lateral tunnel–PAc and between the superior vena cava–PAc were measured in both inspiratory

(patient 7) in class IV (table 2). All patients were in sinus rhythm. Three patients were on anticoagulants and one patient on diuretics (patient 7).

Informed consent was given by patient or parent, as appropriate, and the study was approved by the local ethics committee.

INVESTIGATIONS

On the day of study the patients underwent MR examination immediately before cardiac catheterisation. All patients required sedation (ketamine and propofol) for the catheterisation and four (patients 1, 2, 5, and 6) were also sedated for MR scanning. Heart rate, blood pressure, oxygen saturation, and expired carbon dioxide were recorded continuously in the sedated patients, who were breathing spontaneously and receiving oxygen (1–1.5 l/min) nasally.

Cardiac catheterisation

Standard right and left heart catheterisation was performed via a femoral approach. Pressures were obtained with fluid filled catheters and recorded at 25 mm/s. Biplane angiograms were obtained in the lateral tunnel and in the superior vena cava to assess preferential streaming and diameter changes of the lateral tunnel.

Table 1 Patients' characteristics

Patient	Diagnosis	Connection	Offset	Fenestration	D_{SVC}	D_{LT}	D_{RPA}	D_{LPA}
1	Tricuspid atresia	SVC to RPA, LT to RPA	1 diameter	+	2.0	2.5	1.5	1.5
2	Tricuspid atresia	SVC to RPA, LT to RPA	No offset	+	1.3	–	0.6	0.9
3	Tricuspid atresia	SVC to RPA, LT to RPA	1 diameter	–	1.4	1.6	1.3	1.2
4	Tricuspid atresia	SVC to RPA, LT to RPA	1 diameter	–	1.7	1.8	1.8	1.5
5	Mitral atresia	SVC to RPA, LT to RPA	1 diameter	–	1.1	1.4	1.5	1.3
6	Tricuspid atresia	SVC to RPA, LT to RPA	1 diameter	–	1.3	1.7	1.3	1.0
7	Mitral atresia	LVSC to LPA, LT to RPA	–	+	1.0	1.5	1.1	1.3

D, diameter (cm) calculated as the mean of two perpendicular diameters measured on the MR images. LVSC, left superior vena cava; LT, lateral tunnel; LPA, left pulmonary artery; RPA, right pulmonary artery; SVC, superior vena cava.

Table 2 Flow values

Patient	Q_{SVC}	Q_{LT}	Q_{RPA}	Q_{LPA}	Q_{SVC}/Q_{LT}	Q_{RPA}/Q_{LPA}	Dif%
1*	1.8	2.4	4.3	2.5	0.8	1.7	47
2	1.9	–	1.5	1.2	–	1.2	–
3	1.7	1.4	2.3	1.7	1.3	1.4	25
4	1.2	1.2	1.4	1.0	1.0	1.4	0
5	1.7	1.4	1.7	1.1	1.2	1.5	10
6†	1.5	0.9	1.9	0.8	1.7	2.4	14
7‡	0.6	0.9	0.6	0.8	0.7	0.8	7
Median	1.7	1.3	1.7	1.1	1.1	1.4	12
Mean	1.5	1.4	2.0	1.3	1.1	1.5	17.2
SD	0.5	0.6	1.2	0.6	0.4	0.5	16.8

*No offsetting between the lateral tunnel and the superior vena cava and lateral tunnel flow towards right; †NYHA class III; ‡NYHA class IV, single left superior vena cava, and lateral tunnel flow towards right.
Q, flow (l/min/m²); LPA, left pulmonary artery; LT, lateral tunnel; RPA, right pulmonary artery; SVC, superior vena cava; Dif%, percent difference between inflow and outflow calculated as $((Q_{SVC} + Q_{LT}) - (Q_{RPA} + Q_{LPA})/(0.5 * (Q_{SVC} + Q_{LT}) + (Q_{RPA} + Q_{LPA}))) * 100$.

Table 3 Pulsatility characteristics

Patient	PI_{SVC}	PI_{LT}	PI_{RPA}	PI_{LPA}	dp_{LT}	dp_{SVC}	FS
1*	2.4	8.5	0.8	1.9	4.1	1.6	0.28
2*	0.5	–	1.2	0.9	3.0	1.1	0.13
3	0.8	3.4	0.4	1.6	6.8	2.6	0.18
4	0.5	1.8	1.3	2.0	6.3	1.4	0.13
5	0.9	1.4	1.0	1.7	1.9	1.1	0.09
6†	0.8	2.2	1.9	1.2	–	0.5	0.20
7‡	0.5	1.1	1.4	1.4	4.3	2.5	0.18
Median	0.8	2.0	1.2	1.6	4.2	1.4	0.18

*No offsetting between the lateral tunnel and the superior vena cava and lateral tunnel flow towards right; †NYHA class III; ‡NYHA class IV, single left superior vena cava, and lateral tunnel flow towards right.
PI, pulsatility index; dp_{LT}, maximum instantaneous pressure difference (lateral tunnel (LT) − corresponding pulmonary artery (PAc)) − minimum instantaneous pressure difference (LT − PAc); dp_{SVC}, maximum instantaneous pressure difference (superior vena cava (SVC) − PAc) − minimum instantaneous pressure difference (SVC − PAc); FS, fractional shortening; LPA, left pulmonary artery; RPA, right pulmonary artery.

and expiratory cardiac cycles. Since expiration lasted approximately twice as long as inspiration, mean instantaneous pressures were calculated weighting expiratory values double. From these data, mean respiratory corrected instantaneous pressure difference curves were generated.

The mean gradient across the anastomoses and the maximal (absolute) difference in instantaneous gradients within a cardiac cycle were calculated.

The shortening fraction of the lateral tunnel was calculated as the maximal shortening of the tunnel diameter relative to the maximum dimension halfway between the upper and lower margins in the anterior-posterior projection.

MR scanning
MR data were reconstructed on the MR host computer, and transferred to a SPARC II workstation (Sun Microsystems Inc, Mountain View, California, USA) for artefact compensation.[23] The border of each vessel was manually traced on each time frame. Volumetric flow was calculated for each phase by integration of pixel velocities across the vessel. Flow volume curves were generated and the mean flow was calculated. Flow ratios (right pulmonary artery to left pulmonary artery, and superior vena cava to lateral tunnel) and pulsatility indices for each flow curve were calculated as (maximum flow rate − minimum flow rate)/mean flow rate. To assess the relative contribution to flow by atrial systole, linear interpolation of the flow curve for the missing part of the cardiac cycle was performed.

STATISTICS
Values are given as medians (ranges). Correlations were tested by linear correlation analysis (Spearman). Differences between flow rates and pulsatility indices were tested using Wilcoxon signed rank test. A value of $p < 0.05$ was considered significant.

Results
CARDIAC CATHETERISATION (TABLE 3, FIG 2)
In the three patients (patients 2, 6, and 7) who previously had been palliated with Blalock-Taussig shunts, no pulmonary artery distortion had been caused by the shunt; only in one patient (patient 7) was the upper right lobe vascularisation slightly reduced. In all but two subjects (patients 2 and 7), preferential streaming was from the lateral tunnel to the left pulmonary artery, and from the superior vena cava towards the right pulmonary artery. In the patient with no offsetting between the lateral tunnel and the superior vena cava (patient 2), flow distribution was less lateralised, although flow from the lateral tunnel primarily was towards the right pulmonary artery, and superior vena cava flow predominantly to the left pulmonary artery. In the patient with a left superior vena cava (patient 7), flow from the superior vena cava was mostly directed to the left pulmonary artery, and flow from the lateral tunnel to the right pulmonary artery.

The shortening fraction of the lateral tunnel was 0.18 (0.09–0.28).

The mean pressure gradients across the superior vena cava–PAc and the lateral tunnel–PAc anastomoses were 1.3 (0.7–3.1) mm Hg and 0.6 (0.0–2.0) mm Hg. During the cardiac cycle, the mean instantaneous pressure difference between the lateral tunnel and the PAc was biphasic with two positive peaks, one in late ventricular systole, another in atrial systole. The maximal instantaneous pressure difference between the lateral tunnel and the PAc was significantly higher than the pressure difference between the superior vena cava and the PAc (4.2 (1.1–6.3) *v* 1.5 (1.1–2.53) mm Hg, $p < 0.05$).

MR SCANNING (TABLE 2 AND FIG 2)
The MR flow measurements were complete in all but one patient (patient 2), in whom time restrictions allowed only the left pulmonary artery, right pulmonary artery, and superior vena cava measurements. The sum of volume flow through the superior vena cava and lateral tunnel (3.0 (1.5–4.2) l/min*m²) was similar to volume flow through the right and left pulmonary arteries (2.8 (1.4–6.8) l/min/m² (NS). In one subject (patient 1), the pulmonary outflow, particularly the right pulmonary artery, was significantly higher than caval inflow, possibly because of difficulties in flow measurement caused by the small size of the patient. Overall measurement inaccuracy was 12% (median) (table 2). Right pulmonary artery flow was greater than left pulmonary artery flow (1.7 (0.6–4.3) *v* 1.1 (0.8–2.5) l/min*m², $p < 0.05$) in all but the patient with a left superior vena cava (patient 7). Flow in the superior vena cava
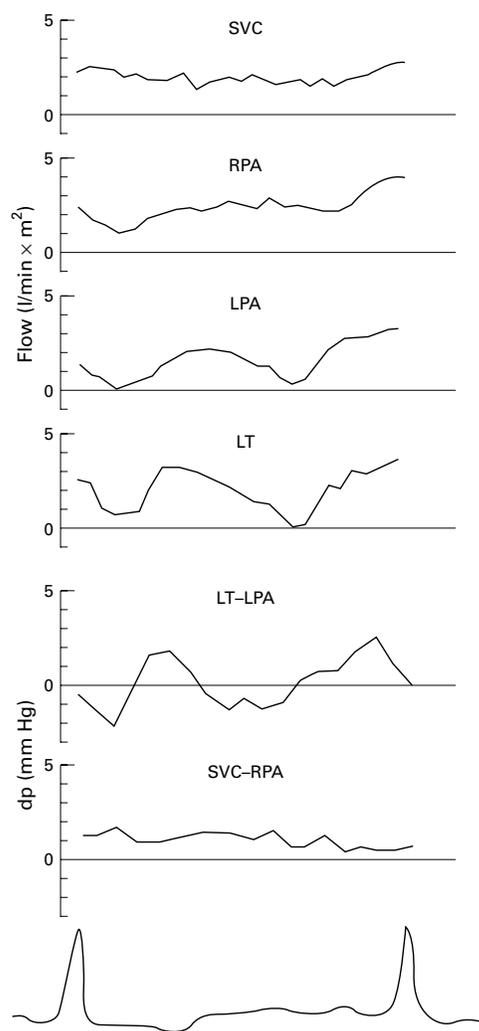
Figure 2   Flow volume curves from the four measurement planes. The instantaneous pressure difference between the lateral tunnel (LT) and the left pulmonary artery (LPA) and between the superior vena cava (SVC) and the right pulmonary artery (RPA) in a typical patient (patient 4). The ECG is shown at the bottom. The flow variation in the lateral tunnel resembles the variations in the pressure gradient between the lateral tunnel and the left pulmonary artery. Flow and pressure curves from the lateral tunnel–left pulmonary artery and from the superior vena cava–right pulmonary artery look similar.

and in the lateral tunnel were similar (1.7 (0.6–1.9) *v* 1.3 (0.9–2.4) l/min⋆m², NS).

PULSATILITY (TABLE 3 AND FIG 2)
The time flow curves in the lateral tunnel revealed a biphasic pulsatile pattern during the cardiac cycle, with a maximum flow rate occurring in atrial systole and a second, but smaller, peak occurring in late ventricular systole. A similar time flow pattern was seen in the PAc—that is, the left pulmonary artery in all but two patients. In contrast, flow in the superior vena cava exhibited only minor temporal variations. The flow curve in the pulmonary artery primarily receiving the superior vena cava flow showed some reflections of the temporal variations seen in the lateral tunnel, but much less than in the contralateral pulmonary artery.

The pulsatility index was higher in the lateral tunnel than in the superior vena cava (2.0

(1.1–8.5) *v* 0.8 (0.4–2.3), p < 0.05). Similarly, the pulsatility index was significantly higher in the pulmonary artery corresponding to the lateral tunnel than in the pulmonary artery corresponding to the superior vena cava (1.6 (1.2–2.0) *v* 1.0 (0.4–1.9), p < 0.05). The pulsatility index in the non-corresponding pulmonary artery was not different from the pulsatility index in the superior vena cava (1.0 (0.4–1.9) *v* 0.8 (0.4–2.3), NS).

There was a positive correlation between the lateral tunnel pulsatility index and the shortening fraction of the lateral tunnel ($r = 0.83$, p < 0.05).

Flow reversal was observed in the lateral tunnel in two patients (patients 1 and 3) but never seen in the pulmonary arteries or in the superior vena cava.

Discussion
After the introduction of the TCPC operation as a final palliative procedure for complex congenital heart malformations,[1] this surgical modification of the Fontan procedure has been widely adopted. However, it is still debated whether the lack of atrial contraction is a major drawback compared with the classic atriopulmonary anastomosis.[10–16] Discussions also relate to the optimal geometry of the venous pathways and anastomoses, including the type of inferior vena cava-superior vena cava pathway (intra- or extra-atrial tunnel), and the optimal mode of cavopulmonary connection such as offsetting, shape, and angulation. These important aspects have mainly been investigated in vitro and by computer simulations, based on assumed input parameters such as inferior vena cava–superior vena cava flow ratios and inflow patterns.[2–6]

This report quantitatively describes the in vivo flow characteristics after the TCPC. Using combined information obtained from MR phase contrast measurements, cardiac catheterisation, and angiography, it was found that right pulmonary artery flow is higher than left pulmonary artery flow, that superior vena cava flow is similar to inferior vena cava (lateral tunnel) flow, and that some pulsatility is preserved in the lateral tunnel and transmitted to both pulmonary arteries, but primarily to the pulmonary artery receiving flow from the lateral tunnel—that is, the left pulmonary artery in most of our patients.

SUPERIOR VENA CAVA–LATERAL TUNNEL (INFERIOR VENA CAVA) FLOW RATIOS AND PULMONARY ARTERY FLOW DISTRIBUTION
In the supine position, the superior vena cava-inferior vena cava ratio of contribution to cardiac output normally falls with increasing age, reaching the adult level of 0.5 at the age of 7 years.[24 25] Despite sedation, which may drop the superior vena cava contribution to cardiac output,[26] the majority of our patients had a superior vena cava–lateral tunnel (inferior vena cava) flow ratio ≥ 1, and none had a flow ratio below 0.7. This suggests that the relative flow distribution from the caval veins is significantly changed after TCPC circulation is established.

The reason for this is unknown but may theoretically be secondary to increased venous resistance in the abdominal organs.[7]

Recent computer simulations and in vitro studies all assume significantly higher flow through the inferior vena cava than through the superior vena cava.[2–6] Although this may be the case during certain types of exercise, our observation of relatively high superior vena cava flow must be considered in simulations and in in vitro studies.

In the normal circulation, flow to the bigger right lung equals or exceeds flow to the smaller left lung.[21 27] In contrast, dominant flow to the left lung has surprisingly been observed after the Fontan operation (atriopulmonary connection).[21] Based on the assumption that approximately two thirds of the systemic venous return comes from the inferior vena cava and mainly goes to the smaller left lung, modification of the originally described TCPC procedure[1] by enlarging the inferior vena cava anastomosis towards the right pulmonary artery has been suggested,[2] although it may be associated with higher energy dissipation.[2] In computer models, this provides increased flow to the right lung and thus an optimal pulmonary flow ratio.[2] Our in vivo MR data, however, clearly show that the standard lateral tunnel TCPC, with lateral tunnel offsetting to the left and superior vena cava offsetting to the right, is associated with a slightly higher right to left pulmonary flow distribution.

PULSATILITY CHARACTERISTICS

The pressure waveform in the lateral tunnel was biphasic with peak pressures occurring in atrial systole and in late ventricular systole. Similar but slightly attenuated pressure waveforms were seen in the pulmonary artery primarily supplied by the lateral tunnel. In contrast, pressure amplitudes in the superior vena cava and its corresponding pulmonary artery were damped. Although the pressure amplitudes may seem small compared to right ventricular amplitudes, the fluctuations compare with normal pulmonary artery pressure pulses, and may thus seem quite sufficient to produce the pulsatility of flow.[28]

Pulsatility was highest in the lateral tunnel and in the pulmonary artery primarily draining the lateral tunnel, and lowest in the superior vena cava. The pulsatility observed in the lateral tunnel was 44% of what has previously been observed in the main pulmonary artery of normal volunteers. In the right and left pulmonary arteries, 30% and 27% of the normal pulsatility in these arteries were seen.[27]

No comparable pulsatility data are available for the atriopulmonary circulation. Rebergen *et al* observed flow reversal in the pulmonary arteries after an atriopulmonary anastomosis.[21] This is in contrast to the present TCPC data but indicates that pulsation is better preserved when the atrium is included in the circulation.

The biphasic pulsation pattern observed in the lateral tunnel, characterised by high flow in atrial systole and late ventricular systole, bears similarities with the v and a waves normally seen in the jugular vein, and has been described

in the pulmonary arteries after an atriopulmonary connection.[21] The observation of similar flow patterns after the TCPC suggests that pressure changes in the left atrium may transmit directly to the lateral tunnel via movements of the patch, as was observed angiographically, or that contraction of the part of the tunnel consisting of atrial wall induces the pulsatility.

Respiration also tends to increase the temporal variations in pressure and flow, producing more dynamic flow in the TCPC circulation than hitherto thought.[29] Thus, semiquantitative MR blood tagging technique has suggested that 30% of flow in the TCPC is respiratory dependent and 70% is cardiac dependent.[30]

The issue of pulsation is of pathophysiological importance because steady flow in the pulmonary arteries may be associated with late pulmonary vascular problems.[10–16] Whether the degree of pulsation observed here is sufficient to protect against the possible deleterious effects of non-pulsatility requires further studies.

LIMITATIONS

Several potential limitations of the present study must be considered.

It must be acknowledged that our data may not necessarily apply to all other TCPC situations. Different types of cavopulmonary anastomoses, presence of fenestrations, type of underlying cardiac malformation, and the overall postoperative result may all influence the postoperative flow dynamics.

Because of heart rate variability and limitations of the MR method used, the last end diastolic part of the cardiac cycle could not be acquired and analysed in all subjects. Linear interpolation, however, suggests that mean flow during this period did not differ significantly from the rest of the cardiac cycle.

Flow characteristics were only studied at rest in the supine position. Although exercise undoubtedly modifies the postoperative flow dynamics, the direction and magnitude of changes associated herewith are unknown, but likely to depend not only on the type of exercise (dynamic *v* static, upper *v* lower limbs) but also on the overall postoperative haemodynamic results.

The importance of respiration on flow dynamics was not addressed, as gating for inspiration and expiration would have quadrupled the examination time.[29] To overcome this limitation, mean values for many cardiac cycles were derived.

CONCLUSIONS

The present study provides quantitative in vivo measurements of flow characteristics after the standard TCPC operation with left offsetting of the inferior vena cava flow and right offsetting of the superior vena cava flow. It confirms that some pulsatility is preserved in the lateral tunnel and to some extent in the pulmonary artery primarily receiving perfusion from the lower part of the body. In the lateral tunnel and the superior vena cava, pulsatile

flow patterns and pressure pulsations are congruent, and in the lateral tunnel the pulsatility index correlated with the shortening fraction.

The flow distribution between the right and left lung is favourable and closer to normal than previously described after an atriopulmonary connection.

These findings are of importance and should be considered in future computer simulations and in vitro studies aiming to define the optimal geometry of the TCPC operation.

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