

A novel technique for non-invasive assessment of pulmonary artery pressure in patients after the Fontan procedure

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Introduction

In the 'Fontan circulation', the caval veins are directly connected to the pulmonary arteries.¹ Due to absence of a subpulmonary ventricle, blood flow through the lungs significantly depends on pulmonary vascular resistance and pulmonary artery pressure (PAP) has an important impact on the outcome of those patients.² The degree of venous congestion is reflected by PAP and central venous pressure (CVP) which are equal in the Fontan physiology³ in the supine but not in the upright position. Evaluation of PAP is of utmost importance as a value > 16 mmHg is considered a predictor of long-term mortality.⁴ However, measurement of PAP is an invasive procedure and thus performed at long intervals only. Therefore, we aimed to analyse the accuracy of a non-invasive method to assess PAP in subjects with Fontan circulation.

Methods

For this prospective study, non-invasive PAP measurements were performed between 01/2024 and 02/2025 at the Pediatric Heart Center Giessen (Germany). We included children and young adults with a total cavopulmonary connection scheduled for cardiac catheterization. Patients with failing Fontan syndrome were excluded from this study.

First, the subjects sat at the edge of the catheterization table in an upright position. Using a linear transducer, the collapse point of the internal jugular vein was located by ultrasound (L12-5 50, Philips, Netherlands, *Figure 1A*) and was marked on the skin. Subsequently, cardiac catheterization was performed in a lying position under sedation with propofol. Invasive PAP (iPAP) at both PAs was recorded. At the end of the procedure, the position of the PAs was marked on the chest under fluoroscopy guidance and the craniocaudal distance between the PAs and the skin mark of the collapse point

of the internal jugular vein was measured (*Figure 1B&C*). This distance was considered the height of the water column over the PAs and the estimated PAP (ePAP) was calculated as: $\text{ePAP (mmHg)} = \text{Distance (cm)} \times 0.74$.

Statistical analysis was conducted using SPSS 29 (IBM SPSS Statistics, USA) and Bland–Altman plots were created.

Results

Nineteen [6/19(32%) female] patients were included in this study. Median age was 17.9 (IQR 11.6–25.8) years. A total of 63% of the subjects (12/19) had a right systemic ventricle. Indication for cardiac catheterization was routine dilatation of already implanted stents in 13/19 (68%) subjects and cyanosis with the objective to identify and occlude veno-venous collaterals in 6/19 (32%) patients.

All patients had a preserved ventricular function, and median B-type natriuretic peptide concentration was 16 (IQR 9–49) pg/mL. On catheterization, median arterial oxygen saturation was 93 (IQR 91–95) %. Median iPAP was 9 (IQR 8.5–10.5) mmHg while ePAP was 10.3 (IQR 9.2–11.8) mmHg. Estimated PAP exceeded iPAP in 13/19 (68%) measurements. *Figure 1D* depicts these results in a Bland–Altman plot. Through the course of the study, the differences between iPAP and ePAP decreased, with a difference < 2 mmHg in the last 10 patients.

Discussion

The present study demonstrates that non-invasive measurement of PAP by localization of the collapse point of the internal jugular vein using ultrasound is feasible in young subjects following the Fontan procedure. Pulmonary artery pressure plays an important role in the follow-up of patients after the Fontan operation as it reflects the degree

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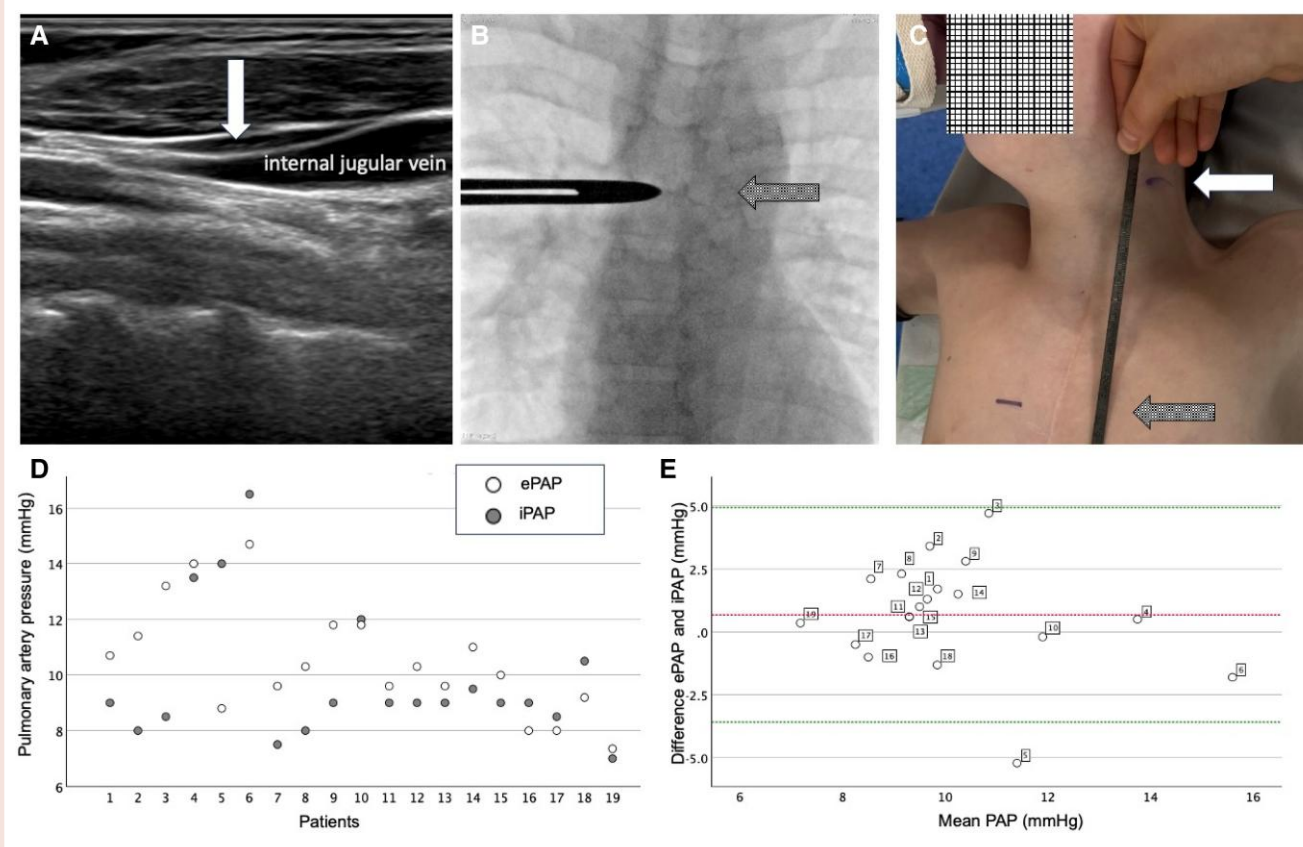


Figure 1 (A) The collapse point of the internal jugular vein was localized by ultrasound (white arrow) in an upright position and was marked on the skin. (B) At the end of catheterization, the level of the pulmonary arteries (PAs) was located by fluoroscopy and was also marked on the skin (grey arrow). (C) The craniocaudal distance between the collapse point of the internal jugular vein and the level of the PAs was measured and used for the estimation of PAP. (D) Estimated non-invasive pulmonary artery pressure (ePAP, open circles) and invasive measurements (iPAP, grey circles) of each participant. Note that ePAP and iPAP did not differ > 2 mmHg during the last 10 measurements. (E) Bland–Altman plot depicting agreement between ePAP and iPAP. The central red dotted line represents the mean difference (0.67 mmHg) between the iPAP and ePAP measurements while the green dotted lines represent the standard deviation. There was only one outlier (Subject No. 5).

of venous congestion before the ‘bottleneck’ of the univentricular physiology which is the blood flow through the lungs.² A high PAP is associated with secondary organ dysfunction like Fontan-associated liver disease or protein-losing enteropathy and is an independent predictor of long-term mortality.⁴ However, measurement of iPAP is resource-intensive, time-consuming, carries risks such as infection, thrombosis, and vascular injury, and requires sedation, particularly in paediatric patients. Efforts have been made to find alternative methods to assess PAP in Fontan patients like measurement of peripheral venous pressure,⁵ ultrasound assessment of the hepatic vein flow pattern,⁶ and diameter of the inferior caval vein or biomarkers⁷ which remain either still invasive or inaccurate.

The technique described in this study is based on the location of the collapse point of the internal jugular vein by ultrasound, and has previously been demonstrated in adult subjects with biventricular physiology to assess CVP.^{8,9} However, our patients were seated upright instead of a 30° position like in healthy controls.⁸ Adjusting the examination position ensures that even elevated PAP can be measured in Fontan patients, as CVP in healthy individuals is normally lower.

The accuracy of this technique was evaluated by comparing non-invasively measured PAP with catheterization data. We found that ePAP was higher than iPAP in 13/19 subjects which is probably caused

by the sedation during the measurement of iPAP leading to lower values. The patient position (sitting at ePAP vs. lying at iPAP assessment) is another important confounder as venous pooling and preload were higher in the horizontal position. Another finding of our study is that the accuracy of ePAP improved with increasing experience leading to PAP differences < 2 mmHg during the last 10 measurements. This improved accuracy was reached by measuring the air-line craniocaudal distance at the ventral aspect of the lying patient despite on the skin surface which avoids miscalculations due to scoliosis (like in Patient No. 5). It is of notice that this technique might be limited to normal or moderately elevated PAP as the sonographic window is limited at the cranial site by the lower edge of the mandible, especially in patients with short status. However, if the collapse point of the jugular vein could not be localized within the sonographic window, a high PAP should be assumed.

Another important factor is the assessment of the PA-level, which served as the ‘lower point’ of the water column in our patients. In this study, the level of the PAs was localized by fluoroscopy which is not feasible in an outpatient setting. Alternatively, the level of the PAs could be located by counting the dorsal ribs as the PAs are commonly situated at the level between the T5 and T6 vertebrae.¹⁰

Overall, non-invasive measurement of PAP provides information about the real-life PAP and appears to be a practical, accurate, and safer alternative to invasive methods in this unique patient population.

Lead author biography



David Backhoff studied medicine at Justus-Liebig University of Giessen and Goethe University of Frankfurt. He trained in Pediatrics at the University Children's Hospital Göttingen, followed by fellowships and board certification in Pediatric Cardiology and Pediatric Intensive Care. Since 2020, he has been a consultant cardiologist at the Department of Pediatric Cardiology, Intensive Care Medicine and Congenital Heart Disease at Justus-Liebig University of Giessen. His clinical and research interests focus on interven-

tional cardiology and electrophysiology in congenital heart disease.

Data availability

Anonymized data of this study are available from the corresponding author upon reasonable request.

Disclosure statement

Consent for the representation of imaging of the patient ([Figure 1C](#)) was given.

Ethical approval

Approved by the local ethical committee.

Funding

None.

Conflict of interest: None declared.

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