

Figure 2. Postnatal CT Angiogram of Case 2

This shows the 5th arch artery connecting the pulmonary trunk and aorta. The communication with the aorta is proximal to the origin of the head and neck vessels. CT = computed tomography.

AP window. The baby was born at term in good condition, and postnatal echocardiography was consistent with prenatal diagnosis. Computed tomography angiography (Fig. 2) was consistent with the remnant of a persistent 5th arch artery, and successful surgery was performed.

The features in case 1 suggested the likely diagnosis of persistent 5th arch rather than an AP window because the AP communicating artery was connecting distal to the origin of the pulmonary arteries. In an AP window, this would be positioned along the pulmonary trunk and bifurcation of the pulmonary arteries. A high AP window may extend to the origin of the right pulmonary artery, but the window is not in the form of a channel with a length resembling a vessel. Moreover, the characteristic wall structure of the arterial duct identified at surgery in our case supported the likely origin of the vessel as a remnant of the fifth arch artery. For the second case, the differentiating point from an arterial duct was the location of the communicating artery connecting the aorta proximal to the origin of the head and neck vessels; an arterial duct would communicate distal to the arch arteries.

Both fetuses reported here likely had vascular channels that represented remnants of a 5th aortic arch artery. Failure to diagnose aortic interruption with persistence of the 5th arch arterial channel in our first case resulted in prolonged surgery and subsequent demise. Lessons learned from this case led to appropriate considerations in the second fetus, confirmation by postnatal imaging, and appropriate surgery that resulted in a good outcome.

Although postnatal reports of a persistent 5th arch artery exist (2), we have demonstrated that fetal diagnosis is possible and that additional imaging, such as computed tomography angiography, is useful to confirm prenatal diagnosis after birth and aid its successful surgical management.

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Repeat Routine Differential Pulmonary Blood Flow Measurements in Congenital Heart Disease by MR

Interstudy Variability and Benchmark of a Clinically Relevant Change

Phase-velocity magnetic resonance (PV-MR) quantifies differential pulmonary blood flow as accurately as the previous gold standard lung perfusion scintigraphy in patients with a single pulmonary blood source supplied by a subpulmonary ventricle (1–3). Therefore, in routine clinical practice, PV-MR has taken on the role of quantifying differential pulmonary blood flow in congenital heart disease (4). However, the interstudy variability and a benchmark of a clinically relevant change of repeat routine differential pulmonary blood flow measurements in congenital heart disease using PV-MR are still unknown.

To test the interstudy variability, we studied 80 consecutive routine clinical cases without intervention or pathological event of the pulmonary branch arteries between 2 consecutive PV-MR measurements (control group). Therefore, in this group, no change of the measured differential pulmonary blood flow ratio was to be expected. This group consisted of 56 cases with repaired tetralogy of Fallot, pulmonary atresia, or truncus arteriosus communis, 7 cases with atrial switch operation, 6 cases with arterial switch operation, 5 cases with Ross operation, and 6 cases with other congenital heart defects. Mean age was 23.2 ± 9.8 years at the time of the first measurement. Mean time between the 2 measurements was 1.2 ± 0.6 years.

To establish a benchmark of a clinically relevant change of repeat routine differential pulmonary blood flow measurements using PV-MR, we studied 13 consecutive routine clinical cases with an explicit unilateral intervention or morphological change to 1 of the pulmonary branch arteries between 2 PV-MR measurements (intervention group). Therefore, in this group, a clear change of the measured differential pulmonary blood flow ratio was to be expected. It is important to note that in our center, PV-MR measurements are not

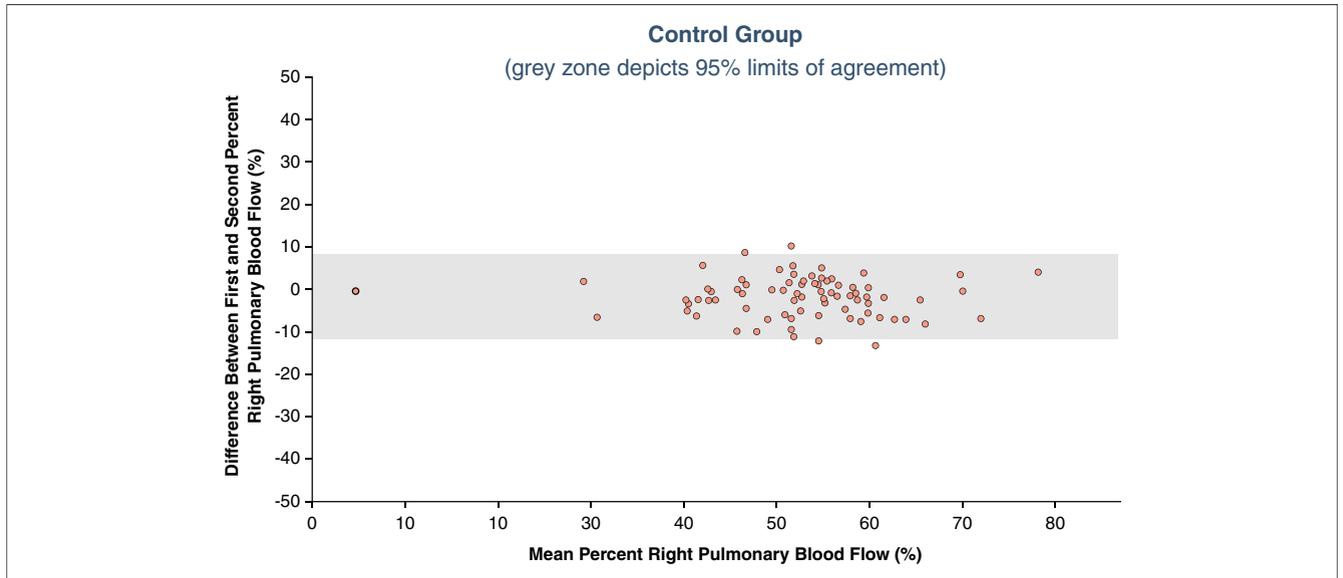


Figure 1. Bland-Altman Analysis of Control Group

Mean difference between 2 consecutive percent right pulmonary blood flow measurements: -2% ; 95% limits of agreement: 8% and -11% (grey zone).

used to decide whether a pulmonary branch artery needs treatment. This group consisted of 11 cases with stent implantation or balloon dilation of 1 of the pulmonary arteries and 2 cases of a post-operative hematoma compressing the left pulmonary artery. Mean age was 21.9 ± 8.3 years at the time of the first measurement. Mean time between the 2 measurements was 0.8 ± 1.0 years.

In each of the measurements, we evaluated net flow volumes in the right and left pulmonary arteries. The representative entity for the differential pulmonary blood flow ratio was defined as percent right pulmonary blood flow as previously described (1) and calculated using the following equation: percent right pulmonary blood

flow = $100 \times [\text{RPA net flow volume}/(\text{RPA net flow volume} + \text{LPA net flow volume})]$, where RPA and LPA are the right and left pulmonary arteries, respectively.

Bland-Altman analysis determined the 95% limits of agreement between 2 consecutive PV-MR measurements (5). The difference between 2 consecutive PV-MR measurements was calculated by subtracting percent right pulmonary blood flow at the first time point from the second time point.

In the control group, the mean difference between the first and second measurements of percent right pulmonary blood flow was -2% with 95% limits of agreement of 8% and -11% (Fig. 1). In contrast to

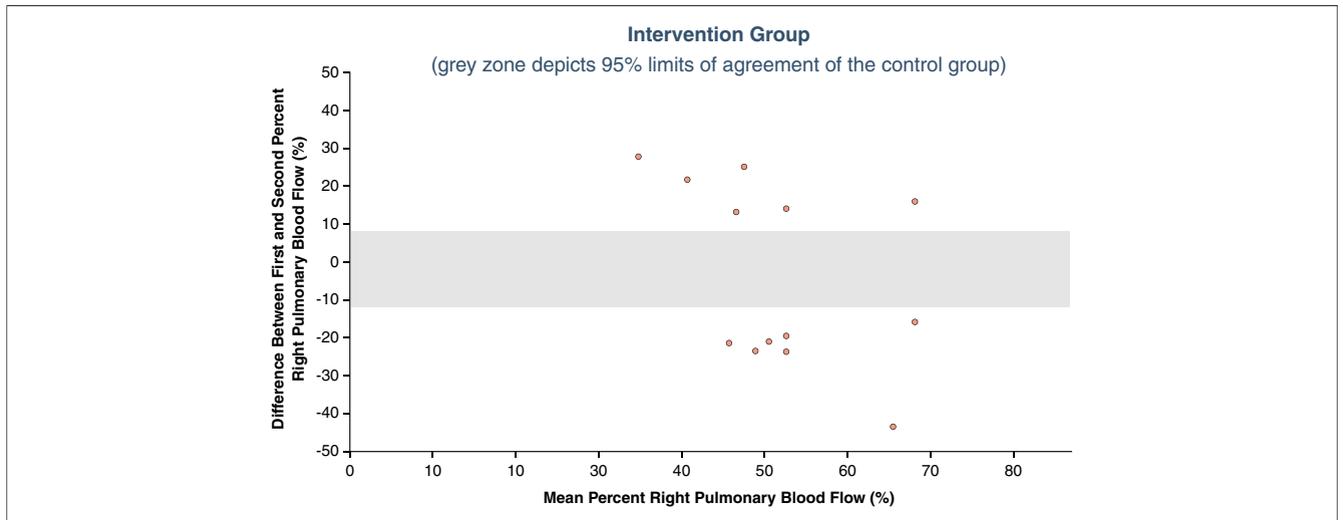


Figure 2. Bland-Altman Analysis of Intervention Group

Mean difference between 2 consecutive percent right pulmonary blood flow measurements: -4% ; 95% limits of agreement: 42% and -50% . Note: all measurements of the intervention group lie outside of the 95% limits of agreement of the control group (grey zone).

this, in the intervention group, the differences between the 2 measurements of percent right pulmonary blood flow were all located outside of the 95% limits of agreement of the control group (Fig. 2).

Furthermore, in the intervention group, the absolute change of percent right pulmonary blood flow was significant at $22 \pm 8\%$ ($p < 0.001$), consistent with the side on which the procedure was performed and the absolute change of the pressure gradient of 22 ± 17 mm Hg ($p = 0.029$).

The results of this study show that consecutive routine clinical differential pulmonary blood flow measurements in congenital heart disease cases without intervention of the pulmonary branch arteries are highly reproducible by PV-MR, with an interstudy variability of up to about 10% (control group, Fig. 1). Moreover, we can use the results of our study to establish a benchmark of a clinically relevant change of repeat routine differential pulmonary blood flow measurements in congenital heart disease using PV-MR in a “real-life” setting (intervention group, Fig. 2). The 95% limits of agreement of repeat routine clinical measurements of percent right pulmonary blood flow by PV-MR of cases without intervention or pathological event of the pulmonary branch arteries (control group) were 8% and –11%. In contrast to this, all measured values in the intervention group were outside of these 95% limits of agreement of the control group. Consequently, we suggest that by using the deviations of 8% and –11% as benchmarks for measurement reproducibility, patients with a clinically relevant change (as assessed by the interventionist) of their differential pulmonary blood flow ratio will correctly be recognized in 100% of the cases. Moreover, patients without

change of their differential pulmonary blood flow ratio will correctly be recognized in 95% of the cases.

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