Unbalanced pulmonary stenosis and pulmonary circulation disturbance persists in patients after Fontan who underwent Norwood-Glenn following bilateral pulmonary banding

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Introduction: In patients with hypoplastic left heart syndrome (HLHS) and the relative disease, we perform bilateral pulmonary artery banding (BPAB) as 1st palliation. As 2nd palliation we previously selected Norwood-Glenn (N-G). On acute stage severe stenosis of left PA frequently occurred. We assessed whether pulmonary circulation disturbance persisted after Fontan in patients who underwent N-G following BPAB. Methods: We defined BPAB group (n=15) as patients who underwent BPAB as 1st palliation, N-G as 2nd palliation, and attained to Fontan procedure. We also defined non-BPAB group (n=72) as patients who got to Fontan through strategy other than BPAB during the same period (between 2007 and 2013). Cardiac catheterization was performed in stable periods after Fontan. The following indexes were calculated with the arterial carbon dioxide pressure and arterial oxygen pressure (PaO2) which were measured during cardiac catheterization: value of alveolar to arterial oxygen tension (AaDO2), RI (AaDO2/PaO2), and P/F (PaO2/0.21). Cardiopulmonary functions were compared between two groups. Results: As for pulmonary circulations diameter of left PA was smaller in BPAB after Fontan (59% vs. 86%, p<0.001). Index of PA was also smaller in BPAB (162 vs. 248 mm/m², p=0.0063). However, there were no significant differences in diameter of right PA. In BPAB AaDO2 was significantly higher (35 vs. 29 mmHg). In BPAB the number of patients with inferior RI (>40) was larger (80% vs. 32%, p <0.001); that with inferior R/F (<322) was larger (40% vs. 13%, p=0.028). Consequently, saturation oxygen of aorta was lower in BPAB (92% vs. 94%, p=0.0077). Smaller major ventricles significantly existed more in BPAB. Other cardiac functions were not significantly different. In BPAB levels of gamma-glutamyl transpeptidase were increased (98 vs. 68U/L, p=0.024), and levels of prothrombin time decayed (67 vs. 74%, p=0.028). Conclusions: Our study showed stenosis of left PA persisted after Fontan in patients who underwent N-G following BPAB. Unilateral stenosis of PA would cause ventilation-perfusion mismatch, which resulted in oxygenation disturbance. Furthermore, considerable smallness of PA might lead to venostasis, which is related to hepatic disturbance. From a long-term perspective on pulmonary circulation, we should select strategy by which pulmonary arteries are grown.