

Frequency of Complications and Outcomes Following Fontan Operation

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Introduction: Fontan operation is the ultimate palliation for CHDs which are not amenable to total correction. This retrospective study investigates the medium and long term complications and outcome of Fontan palliation.

Methods: 90 patients, who underwent Fontan operation between 1987 and 2010 in South Wales, were included in the study.

Results: 52 patients had extra cardiac repair. 46% had fenestration in Fontan circuit. Mean age at operation was 6.1 years (range 2 to 33). Mean follow-up was 7.7 years (range 0 to 26 years). There were a total of 7 deaths with 3 within 30 days of operation, one from PLE, one from arrhythmia, and 2 sudden deaths. The 1-, 5-, 10-, 15, 20 and 25-year survival was 94.9%, 94.9%, 92.5%, 92.5%, 79.3% and 79.3% respectively. In the medium to long term follow-up, 26% had arrhythmias; mostly occurring in the atriopulmonary group 73% versus 13 % in the extracardiac group. 7 patients required permanent pacemaker. 29% had congestive cardiac failure, 7.8% developed protein losing enteropathy (PLE) and 2% developed plastic bronchitis. Two patients with plastic bronchitis made full recovery with PDE5 inhibitor in addition to removal of casts. However only 2/5 patients with PLE responded to medical and catheter/surgical treatment. PLE and plastic bronchitis more often occurred in non-fenestrated group. Congestive cardiac failure occurred more often in the morphological right ventricle group; 73% in the right ventricle group compared to 38% in the left ventricle group were on antifailure medication. There were no thromboembolic complications in any patients who have been on Warfarin.

Conclusions: Arrhythmias and cardiac failure are the most common long-term complications following Fontan procedure. PLE and plastic bronchitis constitute the most challenging clinical problems. Warfarin seems to be effective in preventing thromboembolic events. With growing population of Fontan survivors, efforts to improve Fontan dynamics by supporting cardiac function, or reducing pulmonary vascular resistance should be intensified to minimize the high morbidity and mortality from these complications in the long term.