Protein-Losing Enteropathy after Fontan Procedure

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Objectives: We reviewed the incidence, haemodynamics, treatment strategies and outcome of patients with protein-losing enteropathy (PLE) after Fontan procedure. Methods: The clinical records of 114 patients who underwent Fontan-type operation between 1982 and 1999 were reviewed. The cardiac diagnoses, haemodynamics, clinical presentation, treatment and outcome of those complicated by PLE, as defined by clinical evidence of fluid retention, hypoalbuminaemia (<25 g/L) and enteric loss of protein, were noted. Results: There were 15 early and five late deaths after the Fontan procedure. Three patients defaulted follow-up. Of the remaining 91 patients, five (5.5%) developed PLE. The median age at Fontan operation was 6.4 years (range 1.4 to 15.1). The median age at diagnosis of PLE was 10.9 years (range 6.7 to 19.2), while the median interval between surgery and diagnosis was 4.2 years (range 3 months to 9.5 years). Clinical presentations included oedema (100%), pleural effusion (60%), pericardial effusion (60%), and diarrhoea (20%). Cardiac catheterization revealed an unobstructed Fontan circuit in all of the patients, a mean (± SD) pulmonary arterial pressure of 18±4.8 mmHg (>15 mmHg in three patients), ventricular dysfunction in two patients, and mild prosthetic atrioventricular valve regurgitation in one. Medical treatment with either steroid or heparin resulted in symptomatic improvement in two patients and death in one. Blade atrial septostomy and balloon dilation of the atrial fenestration was performed in two which resulted in normalization of the albumin level in one and death in the other. Conclusion: Protein-losing enteropathy, though relatively uncommon after Fontan operation, is difficult to manage and is associated with morbidity and mortality. An ‘optimal’ post Fontan haemodynamic is not risk free of PLE. (HK J Paediatr (new series) 2002;7:85-91)

Key words: Fontan procedure; Protein-losing enteropathy