We appreciate the comments from D. Simpson that, in addition to intensive medical management for a reduction in central venous pressure (CVP) [1], a drop of oncotic pressure due to hypoalbuminaemia in Fontan patients with protein losing enteropathy (PLE) is another explanation for the absence of difference in the CVP between the ‘excellent’ and chronic phase of PLE Fontan patients [2]. In fact, when we looked into our recent data on consecutive 275 stable Fontan patients, including 21 PLE patients, there were no patients with severe hypoalbuminaemia (≤2.5 g/dl) with a CVP of ≥13 mmHg as Bull [3] noted, although there was a significant inverse correlation between the plasma albumin concentration and CVP (r = –0.23, P < 0.0001).

Pathophysiological mechanisms of hypoalbuminaemia may be multifactorial in Fontan patients with PLE, although the precise mechanism(s) remains unclear. As demonstrated in patients with heart failure, malnutrition, reduced synthesis of albumin in the congested liver with chronic inflammation and increased transcapillary escape rate of albumin may be additional causes of hypoalbuminaemia [4]. We may have to take those into consideration to care for sicker Fontan patients with long-affected PLE because those patients have some pathophysiological conditions similar to those in heart failure patients, such as chronic inflammation [5]. Thus, once PLE occurred, small CVP decline that we observed due to intensive managements and the hypoalbuminaemia-associated low oncotic pressure may not be sufficient to decrease the high rate of albumin transcapillary escape, leading to sustained hypoalbuminaemia, making the situation more susceptible to oedema [6] and intractable to treat. Those sicker PLE patients require repeated hospitalizations, resulting in poor quality of life after the Fontan operation. Our PLE experience reminds us of an importance of selection criteria for the operation and we have to be aware that a role of Fontan operation is to offer a better quality of life in patients with complex congenital heart disease because of its palliative nature.

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REFERENCES