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## Commentary: “A new hope” in patients with Fontan circulation: High-risk Fontan operation can be done, but for “perfect Fontan” we still aim

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A thorough summary of Fontan candidacy and optimization of Fontan circulation was described by Venna and colleagues.<sup>1</sup> They categorized the factors contributing to Fontan indication and late outcomes and proposed a strategy based on the evidence and technologies we currently have. I understood that the authors would like to emphasize the importance of achieving a “perfect Fontan” to be retained as “perfect Fontan circulation” in a lifetime. The strategy they suggested is quite useful and most effective when the problem is the only one and is well documented. However, the real world is more complex and challenging, as most patients with failing or imperfect Fontan circulation have multiple issues. The contributors mentioned by the authors can be divided into 2 factors: ones that appear during the staged repair and the others that occur long-term, after Fontan completion.

For instance, patients with hypoplastic left heart syndrome who are about to undergo Fontan operation may be in excellent condition with good ventricular function and low pulmonary resistance; however, a few patients develop significant tricuspid valve regurgitation with impaired ventricular function and/or poor pulmonary artery growth. We

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### CENTRAL MESSAGE

We can manage the high-risk Fontan operation in the short term but not for a lifetime; hence, we are at the crossroads where we aim for perfect conditions by maximizing the knowledge and technologies.

know that some risk factors developed during the staged repair affect the candidacy for Fontan operation; therefore, treatments to reduce volume loading and aggressive intervention on the atrioventricular valve and pulmonary artery should be planned. Previous studies focusing on long-term outcomes revealed the issues unique to Fontan circulation, such as Fontan-associated liver disease and stenosis of the Fontan pathway. The study showed that the stenting of a narrowed extracardiac conduit was performed well,<sup>1</sup> and this kind of technology is a field that has been constantly growing. In contrast, there remain many risk factors leading to late complications that are not well understood. Recent advancements in surgical techniques and perioperative management have resulted in an unprecedented and excellent short-term outcome after Fontan, but we should realize that we still have ongoing long-term problems that have never been overcome.<sup>2,3</sup> This indicates that the “perfect Fontan candidate” does not guarantee a “perfect Fontan circulation” throughout their entire life.

The authors emphasized that regular exercise is extremely important in patients with a Fontan circulation, to which I completely agree that is true. Although no study has been performed, this phenomenon is often witnessed in clinical practice where the patient’s status suddenly precipitates during hospitalization, as they do not walk like they are at home. Cordina and colleagues<sup>4</sup> reported that lower-limb exercise generates pulsatile flow to the lungs.

Although the exact mechanism has not been elucidated, routine exercise should be recommended for all patients with a Fontan circulation. A game changer such as lower-limb exercise is necessary to obtain the lifelong perfect Fontan as “a new hope.”

Even after recognizing that Fontan circulation is not “perfect,” the indication for Fontan would not be changed in the future. We proceed to Fontan operation in patients even with imperfect conditions; otherwise, quality of life is known to be suboptimal. Our priority is to aim for the “perfect Fontan” using all our knowledge and technologies from the beginning of staged Fontan palliation, as a high-risk Fontan could turn out to be “Darth Vader” later. Together with the evidence for long-term outcomes that were

recently recognized, the treatment should be tailored to manage the “imperfect nature of single ventricle” for the achievement of “perfect Fontan.”

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