

# Case Study: Protein losing enteropathy may pose a hidden danger to prospective heart transplant patients

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## Introduction

Although single antigen and screening beads are highly sensitive and important tools for evaluating pre-transplant compatibility, a patients clinical status or associated treatment often interferes with interpretation. It is important to take such factors into account when making a comprehensive assessment for transplantation. In this case study, we evaluate a patient with protein-losing enteropathy (PLE) in the context of failing Fontan physiology after pediatric palliative treatment for hypoplastic left heart syndrome (HLHS) and the resulting impact to detection of anti-HLA antibody.

## Background

Hypoplastic left heart syndrome is a congenital defect in which the left ventricle, mitral, and aortic valves are underdeveloped or completely closed (fig. 1). Treatment for this condition is palliative and requires a series of three surgeries known as the Norwood, Glenn, and Fontan procedures<sup>1</sup> (fig. 2). Long term, it is possible for cardiac output to decrease to the point of the Fontan procedure being deemed a failure. At this point, the patient may require a heart transplant.

In the Fontan procedure, venous blood is diverted directly to the pulmonary artery, thereby increasing systemic venous pressures<sup>2</sup>. The increased pressure can cause protein to be lost via the intestines (lymphangiectasia) as a means to lower the pressure. This uncompensated loss of plasma protein is a syndrome known as protein-lost enteropathy (PLE) and is acquired in up to 18% of patients undergoing the Fontan procedure<sup>3</sup>. This syndrome presents as nonselective hypoproteinemia with reduced albumin and immunoglobulin levels. PLE can be very mild to severe in patients with elevated systemic venous pressures such that antibody to HLA may not be detectable.

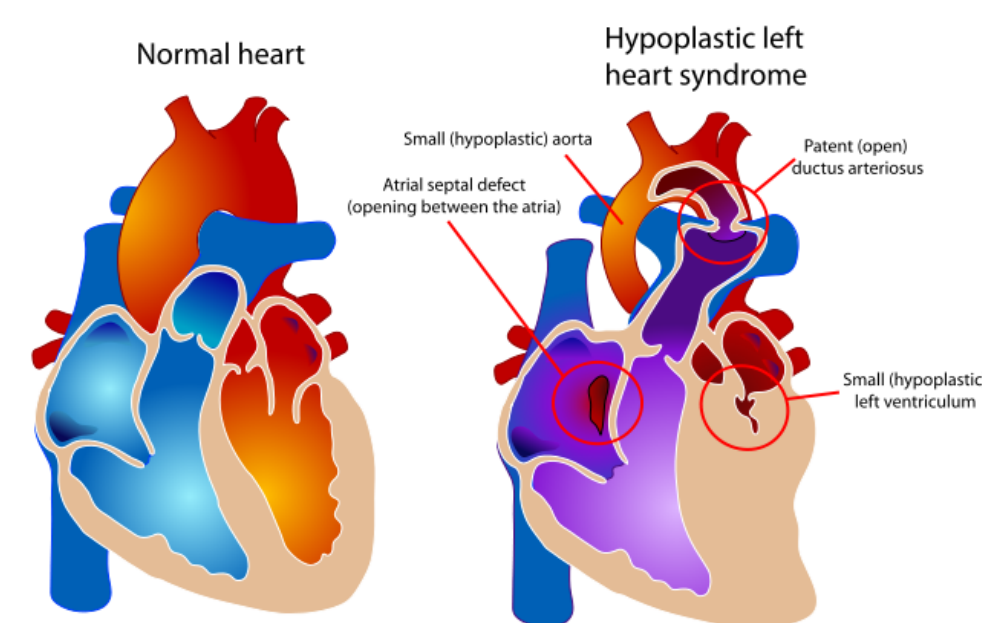


Figure 1. Diagram of Healthy Heart and One with Hypoplastic Left Heart Syndrome<sup>4</sup>.

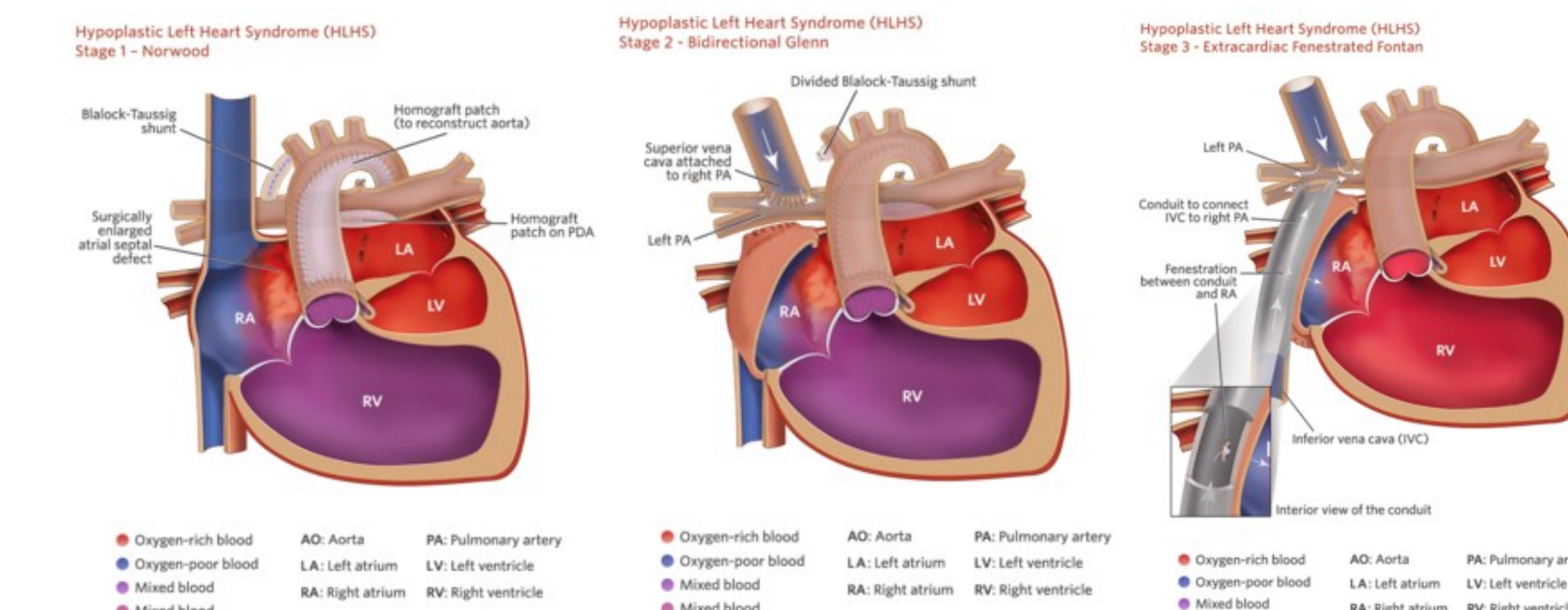


Figure 2. Norwood, Glenn, and Fontan procedures<sup>5</sup>

## Patient Profile

Patient is a 15-year-old male born with HLHS who was palliated with the Fontan procedure. The patient experienced chronic systolic and diastolic heart failure, leading to the need for a heart transplant. As a complication of heart failure and the Fontan procedure, the patient also had longstanding PLE.

## Serum IgG Levels

