# SV-ONE
## Research Explained

Extra-cardiac conduit restriction is associated with increased liver fibrosis in adolescent Fontan patients

Ponzoni M, Saleh J, Chaturvedi RR, Dipchand AI, Valverde I, Seed M, Yoo SJ, Coles J, Honjo O, Lam CZ.J Thorac Cardiovasc Surg. 2025 Oct 3:S0022-5223(25)00855-4. doi: 10.1016/j.jtcvs.2025.09.044. Online ahead of print.PMID: 41047073

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## ABOUT THIS STUDY

The tube (extracardiac conduit or ECC) used to complete the Fontan circuit in the Fontan operation can narrow over time. This study looks at the effects of this narrowing on heart function, exercise tolerance, laboratory tests, and liver fibrosis in Fontan patients.

### Why is this study important?

- Fontan circulation creates a passive, low-pressure system for blood return from the body.
- An ECC does not grow over time. Thus, ECC that is put in early in life – usually during the preschool years - must be large enough function in adulthood.
- Narrowing of the conduit can make it harder for blood to move back to the heart smoothly and efficiently. This increases pressure in various areas of the body – notably the liver.
- Higher pressure in the liver due to the narrowing can lead to irreversible liver disease, including fibrosis (scarring) and/ or cirrhosis – known as Fontan Associated Liver Disease (FALD).
- The size of the ECC typically decreases with time, related to either patient growth and a fixed conduit size or conduit narrowing.
- It is unclear to what extent conduit narrowing contributes to the above issues. However, this could be a target for closer monitoring and intervention.

**How was this study performed?**

- This was a retrospective study looking back as data from March 2018 through October 2022.
- It included 93 adolescent (≤ 18 years) Fontan patients who had an ECC Fontan and underwent a fasting heart-liver MRI scan as part of their usual medical care/surveillance.
- Patients also underwent routine Fontan surveillance testing as part of their usual medical care: heart monitor, blood tests, exercise test, and echocardiogram.
- Conduit size (diameter and area) was measured from the MRI scan.
- The amount of liver fibrosis was graded by how slowly the liver absorbed and released a special contrast dye on the MRI scan. They were then placed into a group 1) none or mild fibrosis or 2) more than mild fibrosis.

**What were the results of the study?**

- A total of 93 patients were included in the study, with an average age of 13.9 years at the time of the MRI. They were an average of 10.2 years out from their ECC Fontan surgery.
- Most patients (94.6%) had a large (20 mm or greater) conduit at the time of the Fontan surgery.
- All 93 patients had conduit measurements available by MRI, and 73% (68/93) of the patients had liver fibrosis grading available by MRI.
- On average there was a decrease in the size of the conduit by 27.3% (6 mm) from the original size.
- A smaller conduit diameter was associated with lower exercise tolerance in patients.
- Patients with complications including protein-loosing enteropathy (PLE), plastic bronchitis, or chylothorax had a significantly smaller conduit diameter compared to those without these conditions.
- A smaller conduit diameter and more years from the Fontan operation were associated with greater than mild liver fibrosis. For every 1 mm larger in diameter the ECC was, the chance of developing greater than mild liver fibrosis decreased by about half.

## What are the limitations of this study

- This was a retrospective design with limitations on the data available for analysis.
- No invasive procedures were performed to obtain additional information, such as liver biopsies or liver-Fontan blood flow gradients in the catheterization lab.
- MRI was the only tool used to assess for liver fibrosis.
- Fontan patients are very complex, and there were only so many variables that were able to be accounted for in the analysis.

## What it all means

- Narrowing of the Fontan ECC narrowing is very common in adolescent Fontan patients and is associated with worse liver fibrosis, PLE, and decreased exercise tolerance.
- Routine MRI surveillance with monitoring of conduit size and liver fibrosis may help identify at-risk patients early on and allow for earlier interventions such as conduit stenting or surgical upsizing, to protect the liver and improve quality of life.
- Further research is needed to explore the long-term benefits of early intervention.