

## Objectives

- The Fontan circulation leads to increased central venous pressures, venous congestion, low cardiac output and provides a predisposed environment for chronic hepatic injury.
- All Fontan patients will develop (some degree) of FALD.
- FALD includes a wide spectrum of pathology that ranges from mild liver fibrosis to liver cirrhosis and hepatocellular carcinoma.
- Optimal modalities for diagnosis and surveillance of FALD are not well defined.

## Methods

- Retrospective review of FALD surveillance data from Fontan patients who had routine follow-up in our department between January 2014 and November 2017
- Surveillance for FALD included liver ultrasound and laboratory testing

### Surveillance for FALD

#### Ultrasound (every 1 – 2 years)

- Surface nodularity
- Heterogeneous parenchyma
- Hyper-echoic lesions
- Portal vein flow (inverse/bidirectional)
- Splenomegaly
- Liver mass
- Ascites

#### Laboratory testing (every year)

- Complete blood count (Platelets)
- AST, ALT (GOT, GPT)
- Gamma-GT
- Bilirubin
- Protein, Albumin
- Prothrombin time (INR)
- Alpha-fetoprotein (AFP)

## Results

- Between January 2014 and November 2017 a total number of 249 Fontan patients had routine follow-up including surveillance for FALD.
- The median age at follow-up was 11 (IQR 7-16) years.
- The median follow-up since Fontan surgery was 8 (IQR 4-13) years.
- 76 (30%) patients had less than 5 years of follow-up, 78 (31%) had a follow-up of 5 to 10 years and 95 (38%) patients had more than 10 years of follow-up.

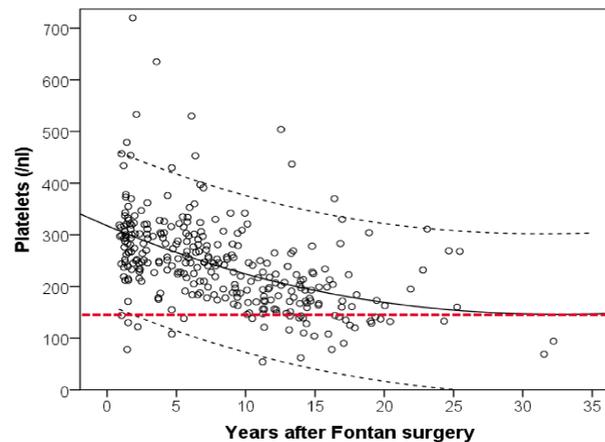


**Figure 3:** Ultrasound images of livers with heterogeneous echotexture showing multiple small sized hyper-echogenic focal lesions with irregular margins. Early stages of fibrotic changes can be best visualized with high-frequency linear probes (3<sup>rd</sup> image).

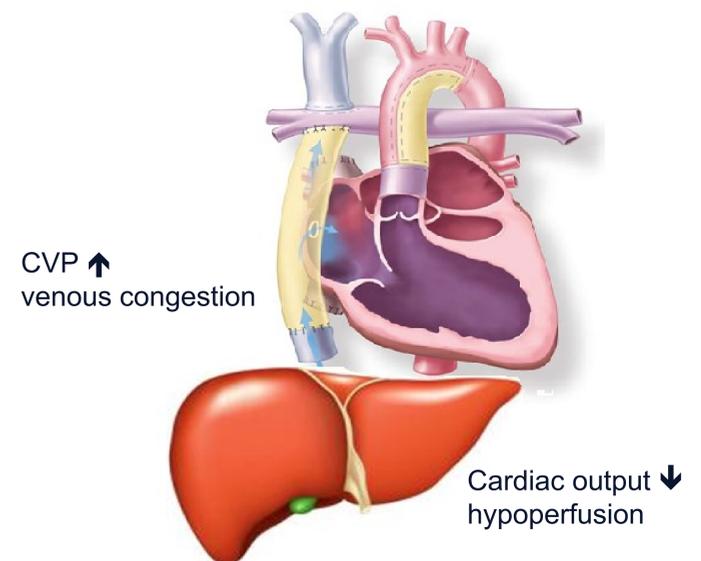
**Figure 4:** The prevalence of fibrotic changes increased with longer follow-up since Fontan completion (<5 years: 25.6%; 5-10 years: 45.5%; >10 years: 58.4%; p=0.004).

	Duration of follow-up		
	< 5 y	5-10 y	> 10 y
Thrombocytopenia (%)	5%	1%	28%
gGT elevated (%)	80%	79%	61%
AST elevated (%)	11%	9%	28%
PT prolonged (%)	22%	33%	29%
Hypoproteinemia (%)	7%	3%	7%
≥ 3 abnormal labs (%)	5%	1%	15%
AST to Platelet Ratio Index	0.3 ± 0.2	0.3 ± 0.2	0.6 ± 0.4
APRI > 0.7 (%)	4%	4%	17%

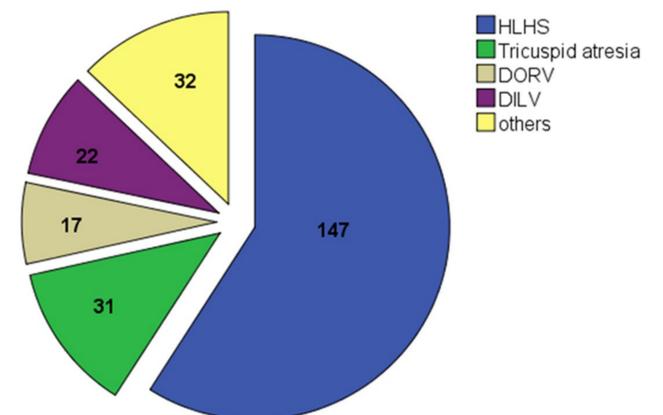
**Table 1:** Thrombocytopenia (platelets <150/nl), AST levels above normal range, the combination of three or more abnormal labs and an AST to Platelet Ratio Index (APRI) above 0.7 were more frequently observed in patients with more than 10 years of follow-up since Fontan surgery.



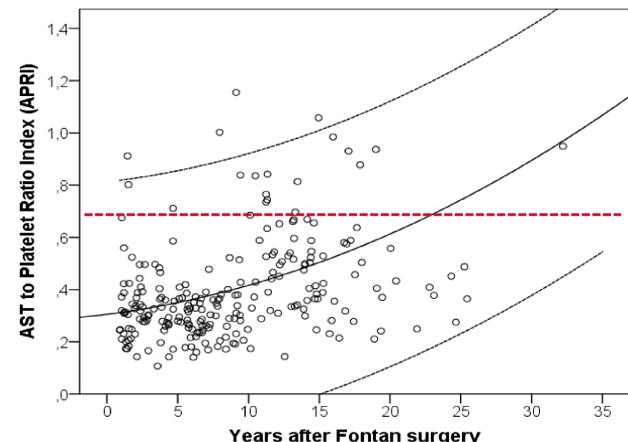
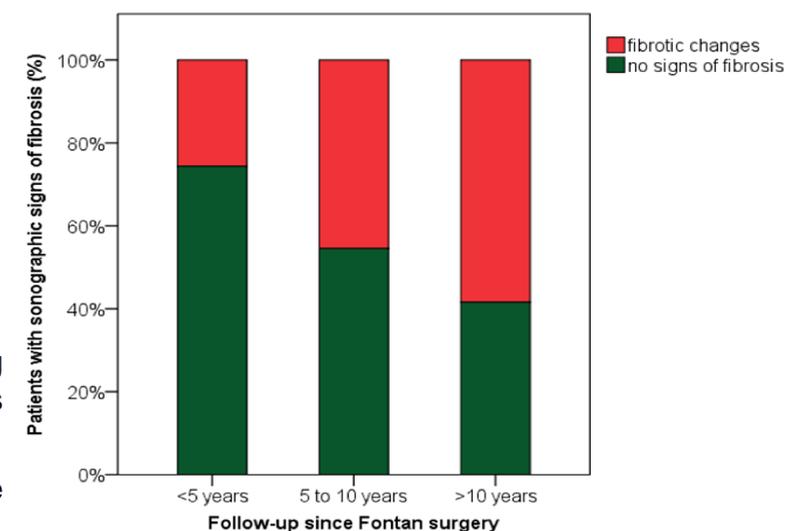
**Figure 5:** The platelet count decreased with longer duration of follow-up after Fontan surgery. There was a moderate correlation between duration of follow-up and platelet count (r=-0.480, p<0.001).



**Figure 1:** The Fontan circulation leads to increased central venous pressures, venous congestion, low cardiac output and thereby to potentially altered end-organ perfusion. The mechanical stress transmitted to the endothelium is one possible mechanisms for the development of fibrosis.



**Figure 2:** Hypoplastic left heart syndrome was the most common diagnosis among Fontan patients screened for FALD.



**Figure 6:** The AST to Platelet Ratio Index (APRI) increased with longer duration of follow-up after the Fontan procedure. There was a only a weak correlation between duration of follow-up and APRI (r=0.373, p<0.001).

## Conclusions

The prevalence of ultrasound and laboratory abnormalities suggestive of FALD increases with longer duration of follow-up after Fontan surgery. Ultrasound abnormalities were not necessary reflected by laboratory abnormalities and vice versa. Further studies are needed to guide surveillance and therapeutic strategies in the management of FALD.