

## MP1-5

### **Percutaneous lymphangiosclerosis as treatment for protein losing enteropathy and plastic bronchitis in patients with failing Fontan circulation.**

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#### **Objectives**

To determine the feasibility and clinical short term results of superselective lymphangiosclerosis in Fontan patients with protein losing enteropathy PLE and plastic bronchitis PB.

#### **Methods**

Dilated lymph vessels in periportal (PLE)(fig A) or paratracheal (PB)(fig B) position were punctured with a 22G Chiba needle; good intralymphatic position ascertained by water soluble contrast injection with drainage to abnormal lacteals; after flushing with glucose 5% (to evacuate most ions), occlusion of distal lymph vessels was obtained by injection of 1-3 cc of a mixture of lipiodol/ n-BCA N-butyl cyanoacrylate (Histoacryl®) 4/1 (which will solidify when in contact with ions as it spreads in lymph vessels). Effect on symptoms, plasma albumin or expectorations was monitored.

#### **Patients & Results**

4 patients with persistent PLE were treated with periportal lymphangiosclerosis; Fontan at  $3.4 \pm 0.4$  years; PLE started  $2.3 \pm 1.0$  (range 1.0-3.3) y after Fontan; time of procedure since start PLE  $8.2 \pm 3.7$  (range 3.3-12.2) y; in all patients (1 patient required a 2<sup>nd</sup> procedure) the lymphangiosclerosis resulted in complete lasting normalisation of albumin levels after withdrawal of all medication (diuretics, steroids, pulmonary vasodilators) (FU 4-8 months). Symptoms of diarrhea and abdominal bloating disappeared with significant improvement of quality of life.

1 pt (Fontan at 2.9 y; age 16.4y) with PB for 2 years had exacerbation of casts expectorations after a surgical procedure. Inguinal intranodal lymphangiography failed to improve symptoms, but demonstrated peritracheal dilated lymphatics. Direct puncture (left and right parasternal) with paratracheal lymphosclerosis resulted in lasting absence of tracheal casts (FU 3 months).

#### **Conclusions:**

Periportal/peritracheal lymphangiosclerosis is a very promising technique in Fontan patients with PLE/PB. Larger series are needed to determine the incidence and reasons of success/failure, with long term results on lymph leak and effects on liver function.

