Introduction:
Pulmonary vein stenosis has an unfavorable outcome because neither surgical nor interventional therapy prevents restenosis. According to promising results in pre-clinical studies, single infants with pulmonary vein stenosis have been treated by balloon dilatation using balloons coated with PACLITAXEL, an antimitotic agent from cancer therapy [1]. First results were encouraging, however, follow-up was cut off early in the two patients published so far, because both died within a few weeks [1,2].

Case Report:
A girl with univentricular heart, increased pulmonary perfusion, and mesocardia was treated by pulmonary banding at 3 weeks. Within the next weeks an increasing stenosis of the left sided pulmonary veins was suspected by echocardiography and confirmed by cardiac catheterization. Subsequently a Damus-Kaye-Stansel anastomosis, an aortopulmonary shunt, and a sutureless repair of the left sided pulmonary venous obstruction were performed at the age of 4 months.

At the age of 6 months, stenosis of the aortopulmonary shunt caused implantation of a 4mm coronary stent. Concurrently severe restenosis of the left pulmonary veins was diagnosed (fig.1) and treated by balloon dilatation.

6 weeks later, re-evaluation in the cath lab revealed severe restenosis, and again dilatation of the left pulmonary veins was performed now using PACLITAXEL coated balloons (5 and 6mm diameter). This procedure was repeated at the age of 10, 13, and 16 months. 2 weeks after the last intervention (fig.2), surgical treatment with right sided Glenn anastomosis and left sided aortopulmonary shunt (5mm) was performed. 8 days after surgery the girl went home.

Out-patient follow-up after 6 weeks revealed the girl in a proper clinical condition with accelerated left-sided pulmonary venous return (Doppler Vmax 2.3m/s). At the age of 22 months the girl was transferred to the cath lab for re-evaluation because of mildly increasing cyanosis. The left sided pulmonary vein showed moderate obstruction, and again re-dilatation was performed using a 6mm PACLITAXEL coated balloon (fig.3).

The right sided Glenn anastomosis was without obstruction, but there was a big anomalous venovenous connection between the superior vena cava and a paravertebral venous plexus draining to the inferior vena cava. The collateral was closed using an Amplatzer duct occluder (fig.4).

Conclusion:
Repeated balloon dilatation of pulmonary venous obstruction using paclitaxel eluting balloons may be useful in the interventional treatment of this frequently fatal condition. Although restenosis occurred also in our patient after the use of paclitaxel eluting balloons, the diameter of the treated vessel showed a reasonable increase, and the patient was able to undergo the next surgical step.

References: