

Therapeutic options and survival of patients with plastic bronchitis after single ventricle palliation – a systematic review

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Introduction: Plastic bronchitis (PB) is a rare complication in single ventricle (SV) patients of which the exact pathophysiology, survival and best treatment are still unclear. This study aims to systematically review the literature to give insight into characteristics, survival and management of SV patients with PB.

Methods: A systematic review was conducted, using the PUBMED database, to find articles, published up to August 2018, with SV patients and PB of which characteristics, treatment and/or outcome were well described per case.

Results: 576 articles were screened and 72 articles had sufficient data of 132 well described SV cases with PB. Most cases had a Fontan palliation (n=125) with a median age at diagnosis of PB of 60.0 months (IQR 41.0-85.3), median age at Fontan operation of 36.5 months (IQR 25.5-50.4), median interval between Fontan operation and diagnosis of PB of 18.0 months (IQR 5.0-36.6) and a median follow-up after diagnosis of PB of 18.0 months (IQR 6.5-36.8). Mortality was 15.2% (n=20) with a median period of 3.5 months after diagnosis of PB. Patients were treated with a combination of medical and interventional/surgical treatment (n=101), only medical (n=13) or only interventional/surgical treatment (n=11). Most reported drugs were fibrinolytics (n=75), most described intervention was bronchoscopic cast extraction (n=61) and most reported catheterization and/or surgical treatments were ligation/embolization of thoracic duct (n=33), relief of arterial, venous or intra-cardiac stenosis (n=26), creation, dilation or stenting of fenestration (n=24), and occasionally Fontan takedown or heart transplantation. Mortality was associated with diagnosis of PB within 12 months after Fontan palliation versus diagnosis after 12 months after Fontan palliation (five-years survival of 56.1% within 12 months vs 94.7% after 12 months, p=0.003; Figure) and a higher age at Fontan operation (47.4 months in the mortality group vs 36.0 months in the survival group, p=0.013).

Conclusions: Most cases are diagnosed with PB one year and a half after the Fontan palliation and around one-sixth of the cases die after a short period after diagnosis. A negative outcome is associated with diagnosis of PB within 12 months after Fontan palliation and a higher age at Fontan operation.

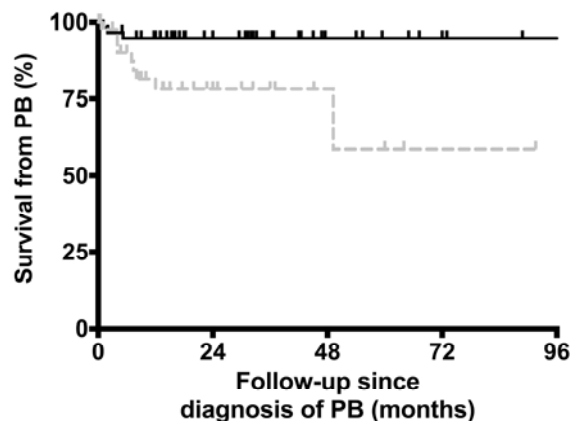


Figure. Kaplan-Meier curve of survival of patients with plastic bronchitis (PB) based on period of diagnosis after Fontan palliation, diagnosis within 12 months (light grey dashed line) and after 12 months (dark grey solid line) after Fontan palliation.