



CONGENITAL LUNG MALFORMATIONS. FROM BABY INTO ADULTHOOD

J.M. Schnater

Pediatric & General Thoracic Surgeon, Assistant professor in Pediatric Surgery, Director of the Pediatric Chest Center, Sophia Children's Hospital, Rotterdam, Netherlands

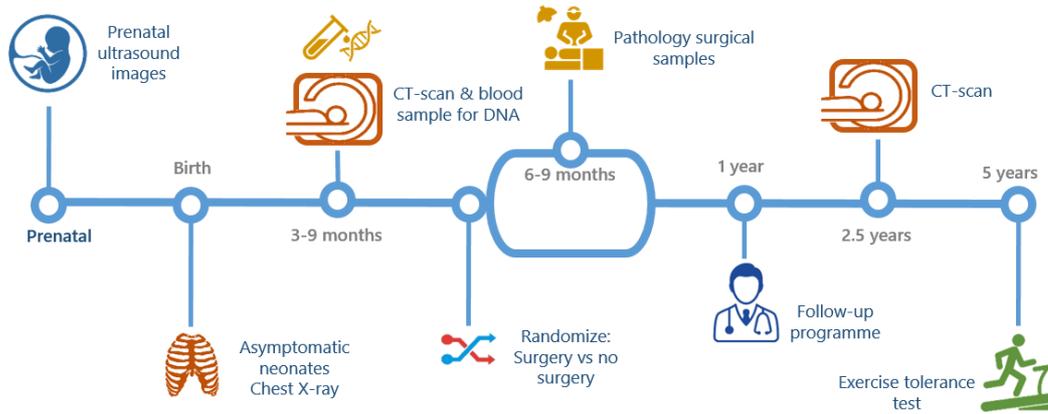
A short overview of congenital lung malformations (CLM) is given, including the natural history and follow-up scheme. The mondial debate about several controversies in CLM is discussed and focus is set on the asymptomatic Congenital Pulmonary Airway Malformation (CPAM) in which consensus on the optimal management is lacking.

The core outcome set for upcoming studies about CPAM is defined during a recently performed DELPHI study in our consortium and the study protocol of the CONNECT trial; the COLlaborative Neonatal Network for the first European CPAM Trial is presented as well.

In this study, patients with a prenatally diagnosed CPAM will be eligible for inclusion and diagnosis will be confirmed with a CT-scan between 3-9 months of age. Simultaneously, blood samples will be obtained for DNA analysis if local legislation permits. After obtaining informed consent, eligible asymptomatic patients will be randomized for either a watchful waiting group or undergo surgical resection between 6-9 months of age. Both groups will be enrolled in a standardized follow-up program with three visits including a repeat CT-scan at 2.5 years and a standardized exercise tolerance test at 5 years (see Figure).

So far, 28 European centres in 16 countries have expressed interest in joining this trial. The primary outcome measure will be the exercise tolerance test at 5 years. Secondary outcome measures will include DNA mutations analysis of DNA, histopathological characteristics, and repeated imaging, as well as pulmonary morbidity and surgical complications during follow-up. By determining predictive features for the development of symptoms, infection, or malignant potential, we aim to identify 'low' and 'high' risk CPAM patients. In our goal working towards a personalised medicine approach.

Figure – trial timeline including the standardised follow-up program



QR-code for details about the CONNECT trial

