

Suspected Bronchiolitis Obliterans, what now?

Dear Parents,

You have received an appointment in our consultation hours for rare lung diseases. We ask you to please bring the following documents to the consultation:

- Insurance card
- The referral letter from your Paediatrician/ General Practitioner
- Any previous diagnostic findings (doctor's letters, results of pulmonary function tests, X-ray/ CT scans and laboratory test results).

We ask you to allow for plenty of time for the medical examination in our outpatient clinic which will include a comprehensive pulmonary function diagnostic.

What is Bronchiolitis obliterans (BO)?

Bronchiolitis obliterans (BO) is a rare and chronic lung disease affecting the lung's tiniest airways, the bronchioles. The bronchioles are the passageways by which air passes through the nose or mouth to the alveoli (air sacs) of the lungs, at which point there is a gas exchange, and oxygen from the alveoli enters into the blood stream. The term Bronchiolitis Obliterans describes the chronic inflammation and scarring of these tiny airways. This leads to a blockage of the airways which may be accompanied by a reduction in lung capacity.

What causes BO?

- Infections
BO can develop following certain viral infections such as infections arising from adenoviruses or atypical infections with mycoplasmas. In some cases it may not be possible to determine whether an infection has caused BO because the triggering bacteria is no longer detectable.
- Transplantations
BO can occur as the result of complications following a bone marrow, stem cell or lung transplantation.
- Bronchopulmonary dysplasia (BPD)
BO can also be a late consequence of premature birth in children.
- Other rare illnesses
BO may also result from other conditions such as connective tissue disorders, acute gastro-esophageal reflux, following smoke intoxication or Stevens-Johnsons Syndrome, a rare reaction to severe infections or specific medication.



Is the diagnosis BO a common one?

No, BO is a rare disease. It is relatively well registered as a complication following transplantations. Not enough data exists identifying BO as a complication following infection. It is therefore not possible to determine how many children and young adults are actually affected in this way.

How is BO diagnosed?

If your doctor suspects that BO is the cause of your child's symptoms, additional testing must be carried to confirm the diagnosis. This will include a comprehensive diagnostic of the pulmonary function and further imaging studies in form of a CT scan.

How is BO treated?

The treatment for BO will start with an anti-inflammatory medication taken orally or by inhalation. There is still no standard treatment plan for BO and therefore each treatment must be adapted individually according to the clinical course of the disease for each patient and discussed and adjusted whenever necessary.

Is there a cure for BO?

This question is difficult to answer based upon the medical knowledge we have today. There are cases where the healing process is able to advance to the stage where the patient experiences a considerable improvement. There are however also cases where the healing process stagnates or leads to a worsening of the condition.

What can parents do?

You, as parents can help your child by being there for them and supporting them as much as you can. You should try to allow for as much normality and daily routine as possible and involve the whole family in supporting the efforts.

You should ensure that you look after yourself too: by seeking support, resting and gathering strength whenever possible and trying not to lose courage.

Where can I obtain more information about BO?

It is not very easy to obtain precise information about BO because it very rare and very little research has been done on the disease.

The first step could be the homepage of the Foundation **Stiftung Starke Lunge**
www.starkelunge.de

There is also a Facebook group in which parents of children with rare lung diseases can exchange information in a closed group www.facebook.com/groups/StärkeLunge.