COUGH ASSIST
(*Mechanical Insufflator-Exsufflator*)

A cough assist (also called a *mechanical insufflator-exsufflator*) is a vital respiratory support tool for people diagnosed with Pompe disease, especially as muscle weakness progresses. Here's how it helps.

Why It's Needed

Pompe disease weakens the diaphragm and other respiratory muscles (including those that control coughing). Because of this, patients often:

- Cannot generate enough force to clear mucus or secretions.
- Are at higher risk for pneumonia, respiratory infections, and lung collapse (atelectasis).

A weak cough means mucus remains in the airways, making it harder to breathe and easier for bacteria to grow.

How the Cough Assist Works

The device helps mimic a natural, strong cough by:

- Inflating the lungs (insufflation) – the device blows air into the lungs to expand them fully.
- Rapidly reversing airflow (exsufflation**)** – it then quickly sucks the air out, simulating the forceful exhalation of a cough.

This cycle can be repeated several times per session to loosen and remove mucus effectively.

Benefits for Pompe Patients

- Improves airway clearance when natural cough is weak.
- Reduces risk of infection and hospitalization.
- Helps maintain better oxygen and carbon dioxide levels.
- May reduce fatigue during illness by lessening the effort needed to clear airways.
- It can be used daily for airway maintenance and during illness for intensive mucus clearance.

When and How It's Used

- Typically prescribed by a pulmonologist familiar with neuromuscular diseases.
- It can be used at home, in hospitals, or in clinics.
- Often combined with other supports such as:
    - Bilevel ventilation (BiPAP) for breathing assistance.
    - Chest physiotherapy or vest therapy to mobilize secretions.
    - Humidification and suctioning if mucus is thick or sticky.

References

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Glossary of Terms:

Airway clearance – The process of removing mucus and secretions from the lungs to keep breathing passages open and reduce the risk of infection.

Atelectasis – Collapse of part or all of a lung, often caused by mucus blocking the airways.

BiPAP (Bilevel Positive Airway Pressure) – A noninvasive breathing machine that gives two air pressures: one to help inhale and one to help exhale. It supports weak breathing muscles.

Chest physiotherapy – A technique that uses manual percussion, vibration, or mechanical devices (like a vest) to loosen mucus in the lungs.

Cough assist (mechanical insufflator-exsufflator) – A machine that helps simulate a natural cough by blowing air into the lungs and then quickly pulling it out to clear mucus when respiratory muscles are too weak to do so.

Diaphragm – The main breathing muscle located under the lungs. Weakness of this muscle makes it hard to inhale fully or cough effectively.

Exsufflation – The “exhale” phase of a cough-assist device, when air is rapidly pulled out of the lungs to mimic a strong cough.

Humidification – Adding moisture to the air from a ventilator or breathing device to prevent airway dryness and thick mucus.

Infection (respiratory) – An illness caused by viruses or bacteria affecting the lungs or airways; often more frequent in people with weak cough or limited airway clearance.

Insufflation – The “inhale” phase of a cough-assist cycle, when air is gently pushed into the lungs to expand them fully.

Lung collapse – See *Atelectasis.* Occurs when a portion of the lung does not fill with air properly.

Mucus / Secretions – The thick fluid produced in the airways that traps dust and germs. In Pompe disease, weak muscles make it difficult to cough mucus out.

Neuromuscular disease – A group of conditions, like Pompe disease, that weaken the muscles controlling movement, breathing, and coughing.

Pulmonologist – A doctor who specializes in lung diseases and respiratory care, often managing cough-assist and BiPAP therapy.

Respiratory muscles – The muscles involved in breathing, including the diaphragm, chest, and abdominal muscles.

Suctioning – The removal of mucus or secretions from the airway using a small tube connected to a suction machine.

Ventilation / Ventilatory support – The movement of air in and out of the lungs. Machines like BiPAP or ventilators provide assistance when breathing muscles are weak.

Vest therapy – Also called “high-frequency chest wall oscillation.” A vibrating vest that loosens mucus to help clear the lungs before or along with cough-assist use.