

People living with Pompe disease commonly develop weakness of the [diaphragm](#) and other respiratory muscles. This leads first to sleep-disordered breathing and nocturnal [hypoventilation](#), and later to daytime [hypercapnia](#). In this context, therapy must provide ventilatory assistance—not just splint the upper airway.

CPAP (Continuous Positive Airway Pressure):

[CPAP](#) delivers a single, constant pressure to keep the upper airway open and is effective for [obstructive sleep apnea \(OSA\)](#). It **does not provide [inspiratory](#) or [expiratory](#) pressure support**, so it does not correct hypoventilation from respiratory muscle weakness and is generally **not appropriate** for Pompe-related hypoventilation unless treating co-existing OSA.

BiPAP / NIV (Bilevel Positive Airway Pressure; Noninvasive Ventilation):

[BiPAP](#) (a form of [NIV](#)) provides **higher inspiratory pressure (IPAP)** and **lower expiratory pressure (EPAP)** to assist [ventilation](#), improve tidal volume, and reduce CO₂ retention. In late-onset Pompe disease, NIV improves nocturnal ventilation, sleep quality, and gas exchange; benefits persist with long-term use.

When to Use NIV in Pompe Disease:

Initiate NIV when clinical features and objective tests indicate evolving hypoventilation or respiratory muscle weakness—for example: symptoms ([orthopnea](#), morning headaches, daytime sleepiness), nocturnal desaturation or CO₂ rise, daytime hypercapnia, or reduced pulmonary function (e.g., markedly reduced [vital capacity](#) or large [supine](#) drop). Decisions should be individualized and guided by polysomnography or overnight oximetry/capnography when available.

Bottom Line:

- **CPAP:** useful for **OSA** but **not** for muscle-weakness-related hypoventilation.
- **BiPAP/NIV:** preferred for **ventilatory support** in Pompe disease and other neuromuscular disorders when hypoventilation is present.

References:

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CPAP VS BIPAP IN POMPE DISEASE

Glossary of Terms:

Airway obstruction – A blockage that prevents air from moving freely in or out of the lungs, often caused by the throat relaxing too much during sleep.

BiPAP (Bilevel Positive Airway Pressure) – A type of noninvasive ventilator that gives two levels of air pressure: one for breathing in (to help fill the lungs) and a lower one for breathing out (to make exhaling easier).

CPAP (Continuous Positive Airway Pressure) – A simpler device that provides one steady air pressure to keep the airway open, mainly used for sleep apnea but not for weak breathing muscles.

Diaphragm – The main muscle under the lungs that helps draw air in when you breathe; it can weaken in Pompe disease.

Expiratory – Refers to exhalation, or the act of breathing air out of the lungs.

FVC (Forced Vital Capacity) – A breathing test that measures how much air you can blow out after taking a deep breath. It helps doctors check lung strength.

Hypercapnia – A build-up of carbon dioxide (CO₂) in the blood, which can cause morning headaches, sleepiness, and fatigue.

Hypoventilation – Breathing that is too shallow or slow to remove enough carbon dioxide from the body.

Inspiratory – Refers to inhalation, or the act of breathing air into the lungs.

Noninvasive Ventilation (NIV) – Any form of breathing support that uses a mask instead of a breathing tube to help air move in and out of the lungs.

Orthopnea – Shortness of breath that happens when lying flat; sitting up usually makes breathing easier.

OSA (Obstructive Sleep Apnea) – A condition where the upper airway repeatedly collapses during sleep, causing pauses in breathing.

Supine position – Lying flat on your back; breathing can be harder in this position if your diaphragm is weak.



CPAP VS BIPAP IN POMPE DISEASE

Ventilation – The movement of air in and out of the lungs; when the muscles are weak, machines like BiPAP can help maintain it.