MY ACHON JOURNEY
Pathways to Navigating Achondroplasia

The guide is not intended to serve as medical advice or to substitute for consultation with a US health care professional. Always talk to your doctor if you have any health care related questions or concerns.

Supplemental oxygen use in infants and toddlers with achondroplasia

Why would a child with achondroplasia need supplemental oxygen?

A child with achondroplasia may be treated with **supplemental oxygen** if they have breathing challenges. The most common cause of breathing difficulty in children with achondroplasia is sleep apnea, a condition in which breathing repeatedly stops and starts during sleep. Supplemental oxygen can help maintain a safe level of oxygen in their body. Oxygen is delivered to the child from a metal tank or machine (*concentrator*) through a plastic tube (*nasal cannula*) placed at the base of the nose. This guide covers the basics for using supplemental oxygen.

Instructions for home oxygen use may vary based on equipment manufacturers and individual medical needs. Always follow the instructions provided by the child's oxygen supply company and/or health care provider.

Child's oxygen prescription:

Frequency of oxygen therapy (choose one): ☐ **All the time** ☐ **During sleep only**

Type of oxygen flow (choose one): ☐ **Continuous** ☐ **Pulse**

Oxygen flow rate: _____ **liters per minute**

The flow rate is the child's prescription and should not be adjusted without consulting their doctor.

If you have questions about supplemental oxygen therapy, contact the child's care provider or oxygen supply company.

Care provider

Daytime: _____

After-hours: _____

Oxygen supply company

Daytime: _____

After-hours: _____

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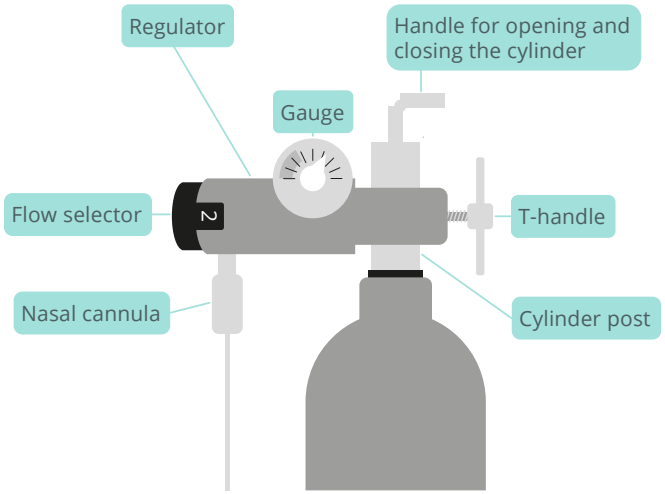
Using an oxygen tank

An **oxygen tank** is a metal tank that holds oxygen. These tanks come in large and small sizes that can be used at home or on the go. To use an oxygen tank:

1. Wash your hands
2. Attach the regulator to the oxygen tank
   - Loosen the T-handle on the regulator
   - Lower the regulator onto the cylinder post. Make sure the pins on the regulator match up with the holes on the cylinder post
   - Tighten the T-handle to secure the regulator to the cylinder post
3. Open the cylinder by turning the top handle counter-clockwise (*"lefty-loosey"*)
4. Check the gauge to make sure there is enough oxygen in the tank
5. Attach the cannula tubing to the regulator. Make sure there are no kinks or blockages in the tubing and check that oxygen is flowing out of the prongs (using a clean hand)
6. Attach the nasal cannula to the child's face (see *How to put on the nasal cannula*)
7. Set the flow selector to the prescribed flow rate (see *Child's oxygen prescription*)
8. While in use, the oxygen tank should be secured upright in a carrier or lying flat on the ground
9. When the child is done using the oxygen tank, turn the cylinder off by turning the handle clockwise (*"righty-tighty"*) and turn the flow selector to the "zero" position

When the tank runs out of oxygen, new tanks can be delivered by the oxygen supply company, or the tank can be refilled with a home fill system according to the instructions provided by the oxygen supply company.

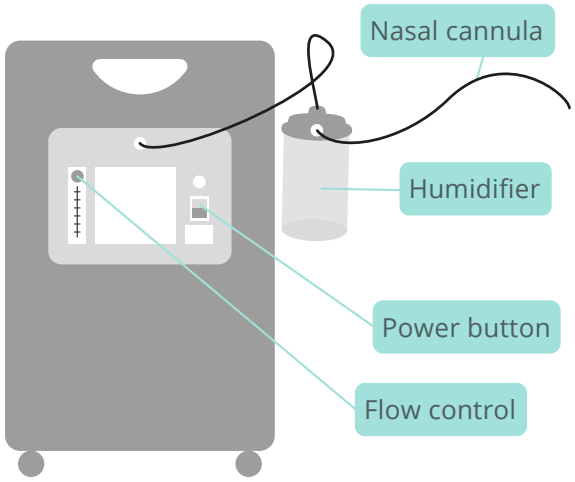


Oxygen tank with regulator attached

Using an oxygen concentrator

An **oxygen concentrator** takes air from your surroundings, pulls out the oxygen, and filters it for the child to breathe. Oxygen concentrators are typically used at home, although portable versions may be used on the go. To use an oxygen concentrator:

1. Fill the humidifier bottle with distilled water and attach it to the oxygen outlet (if applicable)
2. Make sure the concentrator is plugged into a grounded wall outlet and placed 1 to 2 feet away from any wall
3. Attach the cannula tubing to the concentrator. Make sure there are no kinks or blockages in the tubing and check that oxygen is flowing out of the prongs (using a clean hand)
4. Turn on the concentrator power
5. Adjust the flow control to the prescribed flow rate (see *Child's oxygen prescription*)
6. Check for alarms that may indicate a problem, such as power loss or no oxygen flow (refer to the instructions provided by the oxygen supply company for more information)
7. Attach the nasal cannula to child's face (see *How to put on the nasal cannula*)



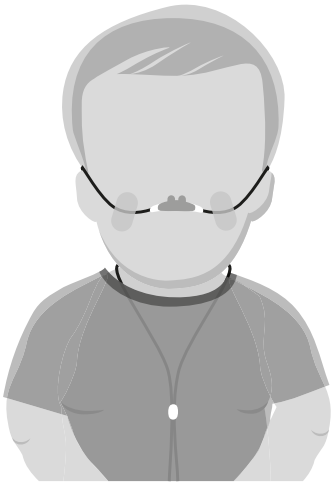
Oxygen concentrator

Placing a nasal cannula

The **nasal cannula** is the plastic tubing through which oxygen is delivered to the child. It has two prongs that go inside their nostrils. To place the nasal cannula on the child's face:

1. Make sure their face is clean and dry
2. Apply a skin barrier sticker (*hydrocolloid dressing*) on each cheek to protect from irritation
3. Secure the tubing to their face using the adhesives provided by the oxygen supply company
4. Tuck the tubing behind their ears
5. Bring the tubing down the front of their body underneath their clothing and allow it to exit from an opening in their clothing (outfits with open feet or double-zippers can be helpful)
6. Make sure there are no kinks or blockages in the tubing
7. After the cannula is attached to their face, connect the other end to the oxygen delivery device (please see *Using an oxygen tank* or *Using an oxygen concentrator*)

To prevent the inside of the child's nose from becoming dry, clean their nostrils with a soft, moist cloth and apply a water-based lubricant where the cannula rests.



A nasal cannula secured to a child's face

Using a pulse oximeter

A **pulse oximeter** (or "pulse ox") is an electronic device that measures the amount of oxygen in the blood to make sure it does not fall to a dangerously low level. To use a pulse oximeter:

1. Following the instructions provided by the oxygen supply company, attach the probe to the child's big toe or finger
2. Wait a few seconds for the pulse oximeter to register the child's oxygen level and heart rate

A typical oxygen level for a baby is between 95% and 100%. If the reading is lower than 95%, consult a health care professional promptly. Note that movement can cause false alarms, so the child must be sitting or lying still to get an accurate reading.

Safety

- Do not allow smoking, open flames, flammable products (such as petroleum jelly, hand sanitizer, rubbing alcohol, and aerosol sprays), or electrical or spark hazards in the room or area where the oxygen equipment is located
- Keep oxygen tubing away from walking areas where it may cause someone to trip and fall
- Avoid leaving oxygen tanks in a hot car
- **If the child is unresponsive and is not breathing, call 911 and follow their instructions**

References:
1. Pauli R, et al. *Orphanet J Rare Dis*. 2019;14(1):1-49. **2.** Hoover-Fong J, et al. *Pediatrics*. 2020;145(6):e20201010. **3.** Afsharpaiman S, et al. *Paediatr Respir Rev*. 2013;14(4):250–255. **4.** Okenfuss E, et al. *Am J Med Genet*. 182A:2540–2551. **5.** American Lung Society. https://www.lung.org/lung-health-diseases/lung-procedures-and-tests/oxygen-therapy. Accessed 2 April 2024. **6.** American Thoracic Society. *Am J Respir Crit Care Med*. 2019;199:5–6. **7.** MedPro Respiratory Care. *Regulator Instructions*. 2013:1–8. **8.** Oxygen Concentrator Store. https://www.oxygenconcentratorstore.com/help-center/how-to-operate-an-oxygen-concentrator/. Accessed 2 April 2024. **9.** Healthwise. https://www.myactivehealth.com/hwcontent/content/special/ut1104.html. Accessed 2 April 2024. **10.** Hy-Tape. https://hytape.com/neonatal-skin/tips-for-securing-the-cannula-in-infants-and-children. Accessed 2 April 2024. **11.** Women's and Children's Health Network. https://cdn.wchn.sa.gov.au/downloads/WCH/hospital-services/home-oxygen/oxygen-cannula.pdf. Accessed 2 April 2024. **12.** Coping with Laryngomalacia, Inc. https://www.copingwithlm.org/product-page/oxygen-feeding-tube-friendly-pajamas. Accessed 20 March 2024. **13.** Lepu Medical. https://en.lepumedical.com/how-to-use-pulse-oximeter-on-baby.html. Accessed 2 April 2024. **14.** Balfour-Lynn M, et al. *Thorax*. 2005;60:76–81. **15.** Drugs.com. https://www.drugs.com/cg/oxygen-therapy-in-neonates.html. Accessed 2 April 2024.

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