

Oxygen Use for Individuals with Cystinosis with Severe Muscle Weakness

As people with cystinosis get older, muscle strength can decrease and affect how well they breathe. Those with more severe muscle weakness may benefit from breathing supplemental oxygen, particularly if they have additional lung conditions. The decision whether someone needs oxygen depends on the amount of oxygen that is present in arterial blood. The simplest way to determine this is by a pulse oximeter that measures the percent of hemoglobin in blood that is saturated with oxygen. Generally, a level of 89% saturation or above is considered adequate and supplemental oxygen is not needed. However if it is below this level, it is appropriate for a physician to prescribe supplemental oxygen for home use. In general, insurance companies will cover the cost of home oxygen when oxygen saturation is 88% or below. Some people may have adequate oxygen saturation when they are at rest but it drops with exercise or during sleep. For these people, supplement oxygen may be beneficial when they are physically active or when sleeping. It may require an exercise test or a sleep study to determine this.

Muscle weakness that affects breathing (restrictive lung disease) may cause problems during chest infection such as pneumonia. The muscles may not be strong enough to allow for comfortable breathing or for someone to cough out secretions from the airways. Treatment of pneumonia requires prompt use of antibiotics. Devices used to assist breathing such as BiPAP may also be needed. Depending on the circumstances, supplemental oxygen may be an important part of the treatment. Again, the decision whether supplemental oxygen will help is based on the oxygen saturation level of blood. Patients are usually given supplemental oxygen to keep the saturation level above 88%. The oxygen treatment is continued until the patient recovers and the oxygen saturation level increases to above 88%.

Questions about oxygen use should be directed to your primary care provider and/or your pulmonologist.

It is highly recommended that patients with cystinosis undergo pulmonary function testing and have regular pulmonary follow-up if they have any significant muscle weakness and particularly if they become short of breath during activities that previously they could do without difficulty .

Vaccinations against influenza (injectable form, not live intranasal) and pneumococcus (Pneumovax, Prevnar) should be discussed with your primary care provider/transplant nephrologist/pulmonologist.

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