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103.00 Respiratory System - Childhood

Section 103.00 Respiratory System

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- 103.01** [Category of Impairments, Respiratory System](#) A. *Introduction.* The listings in this section describe impairments resulting from respiratory disorders based on symptoms, physical signs, laboratory test abnormalities, and response to a regimen of treatment prescribed by a treating source. Respiratory disorders, along with any associated impairment(s) must be established by medical evidence. Evidence must be provided in sufficient detail to permit an independent reviewer to evaluate the severity of the impairment. Reasonable efforts should be made to ensure evaluation by a program physician specializing in childhood respiratory impairments or a qualified pediatrician.
- 103.02** [Chronic pulmonary insufficiency](#)
- 103.03** [Asthma](#) Many children, especially those who have listing-level impairments, will have received the benefit of medically prescribed treatment. Whenever there is such evidence, the longitudinal clinical record must include a description of the treatment prescribed by the treating source and response, in addition to information about the nature and severity of the impairment. It is important to document any prescribed treatment and response because this medical management may have improved the child's functional status. The longitudinal record should provide information regarding functional recovery, if any.
- 103.04** [Cystic Fibrosis](#)
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Some children will not have received ongoing treatment or have an ongoing relationship with the medical community, despite the existence of a severe impairment(s). A child who does not receive treatment may or may not be able to show an impairment that meets the criteria of these listings. Even if a child does not show that his or her impairment meets the criteria of these listings, the child may have an impairment(s) that medically or functionally equals the listings.

Unless the claim can be decided favorably on the basis of the current evidence, a longitudinal record is still important because it will provide information about such things as the ongoing medical severity of the impairment, the level of the child's functioning, and the frequency, severity, and duration of symptoms. Also, the asthma listing specifically includes a requirement for continuing signs and symptoms despite a regimen of prescribed treatment.

Evaluation should include consideration of adverse effects of respiratory impairment in all relevant body systems, and especially on the child's growth and development or mental functioning, as described under the growth impairment (100.00), neurological (111.00), and mental disorders (112.00) listings.

It must be remembered that these listings are only examples of common respiratory disorders that are severe enough to find a child disabled. When a child has a medically determinable impairment that is not listed, an impairment that does not meet the requirements of a listing, or a combination of impairments no one of which meets the requirements of a listing, we will make a determination whether the child's impairment(s) medically or functionally equals the listings. (See §§ 404.1526, 416.926, and 416.926a.)

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B. Documentation of Pulmonary Function Testing. The results of spirometry that are used for adjudication, under the 103.02 A and B, 103.03, and 103.04 of these listings should be expressed in liters (L), body temperature and pressure saturated with water vapor (BTPS). The reported one-second forced expiratory volume (FEV_1) and forced vital capacity (FVC) should represent the largest of at least three satisfactory forced expiratory maneuvers. Two of the satisfactory spirometry tests should be reproducible for both pre-bronchodilator tests and, if indicated, postbronchodilator tests. A value is considered reproducible if it does not differ from the largest value by more than 5 percent or 0.1 L, whichever is greater. The highest values of the FEV_1 and FVC, whether from the same or different tracings, should be used to assess the severity of the respiratory impairment.

Peak flow should be achieved early in expiration, and the spirometry test should have a smooth contour with gradually decreasing flow throughout expiration. The zero time for measurement of the FEV_1 and FVC, if not distinct, should be derived by linear back-extrapolation of peak flow to zero volume. A spirometry test is satisfactory for measurement of the FEV_1 if the expiratory volume at the back-extrapolated zero time is less than 5 percent of the FVC or 0.1 L, whichever is greater. The spirometry test is satisfactory for measurement of the FVC if maximal expiratory effort continues for at least 6 seconds, or if there is a plateau in the volume-time curve with no detectable change in expired volume (VE) during the last 2 seconds of maximal expiratory effort.

Spirometry should be repeated after administration of an aerosolized bronchodilator under supervision of the testing personnel if the prebronchodilator FEV_1 value is less than the appropriate reference value in table I or III, as appropriate. If

a bronchodilator is not administered, the reason should be clearly stated in the report. Pulmonary function studies should not be performed unless the clinical status is stable (e.g., the child is not having an asthmatic attack or suffering from an acute respiratory infection or other chronic illness.). Wheezing is common in asthma, chronic bronchitis, or chronic obstructive pulmonary disease and does not preclude testing.

Pulmonary function studies performed to assess airflow obstruction without testing after bronchodilators cannot be used to assess levels of impairment in the range that prevents a child from performing age appropriate activities unless the use of bronchodilators is contraindicated. Post-bronchodilator testing, should be performed 10 minutes after bronchodilator administration. The dose and name of the bronchodilator administered should be specified. The values in 103.02 and 103.04 must only be used as criteria for the level of ventilatory impairment that exists during the child's most stable state of health (i.e., any period in time except during or shortly after an exacerbation).

The appropriately labeled spirometric tracing, showing the child's name, date of testing, distance per second on the abscissa and the distance per liter (L) on the ordinate, must be incorporated into the file. The manufacturer and model number of the device used to measure and record the spirogram should be stated. The testing device must accurately measure both time and volume, the latter to within 1 percent of a 3 L calibrating volume. If the spirogram was generated by any means other than direct pen linkage to a mechanical displacement-type spirometer, the testing device must have had a recorded calibration performed previously on the day of the spirometric measurement.

If the spirometer directly measures flow, and volume is derived by electronic integration, the linearity of the device must be documented by recording volume calibrations at three different flow rates of approximately 30 L/min (3 L/6 sec), 60 L/min (3 L/3 sec), and 180 L/min (3 L/sec). The volume calibrations should agree to within 1 percent of a 3 L calibrating volume. The proximity of the flow sensor to the child should be noted, and it should be stated whether or not a BTPS correction factor was used for the calibration recordings and for the child's actual spiograms.

The spirogram must be recorded at a speed of at least 20 mm/sec, and the recording device must provide a volume excursion of at least 10 mm/L. If reproductions of the original spirometric tracings are submitted, they must be legible and have a time scale of at least 20 mm/sec and a volume scale of at least 10 mm/L to permit independent measurements. Calculation of FEV_1 from a flow-volume tracing is not acceptable; i.e., the spirogram and calibrations must be presented in a volume-time format at a speed of at least 20 mm/sec and a volume excursion of at least 10 mm/L to permit

independent evaluation.

A statement should be made in the pulmonary function test report of the child's ability to understand directions, as well as his or her effort and cooperation in performing the pulmonary function tests.

Purchase of a pulmonary function test is appropriate only when the child is capable of performing reproducible forced expiratory maneuvers. This capability usually occurs around age 6. Purchase of a pulmonary function test may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided.

The pulmonary function tables in 103.02 and 103.04 are based on measurement of standing height without shoes. If a child has marked spinal deformities (e.g. kyphoscoliosis), the measured span between the fingertips with the upper extremities abducted 90 degrees should be substituted for height when this measurement is greater than the standing height without shoes.

C. Documentation of chronic impairment of gas exchange.

1. Arterial blood gas studies (ABGS). An ABGS performed at rest (while breathing room air, awake and sitting or standing) should be analyzed in a laboratory certified by a State or Federal agency. If the laboratory is not certified, it must submit evidence of participation in a national proficiency testing program as well as acceptable quality control at the time of testing. The report should include the altitude of the facility and the barometric pressure on the date of analysis.

Purchase of a resting ABGS may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided. Before purchasing resting ABGS a program physician, preferably one experienced in the care of children with pulmonary disease, must review the clinical and laboratory data short of this procedure, including spirometry, to determine whether obtaining the test would present a significant risk to the child.

2. *Oximetry*. Pulse oximetry may be substituted for arterial blood gases in children under 12 years of age. The oximetry unit should employ the basic technology of spectrophotometric plethysmography as described in Taylor, M.B., and Whitwain, J.G., "Current Status of Pulse Oximetry," "Anesthesia," Vol. 41. No. 9, pp. 943-949, 1986. The unit should provide a visual display of the pulse signal and the corresponding oxygen saturation. A hard copy of the readings (heart rate and saturation) should be provided. Readings should be obtained for a minimum of 5 minutes. The written

report should describe patient activity during the recording; i.e., sleep rate, feeding, or exercise. Correlation between the actual heart rate determined by a trained observer and that displayed by the oximeter should be provided. A statement should be made in the report of the child's effort and cooperation during the test.

Purchase of oximetry may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided.

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D. *Cystic fibrosis* is a disorder that affects either the respiratory or digestive body systems or both and may impact on a child's growth and development. It is responsible for a wide and variable spectrum of clinical manifestations and complications. Confirmation of the diagnosis is based upon an elevated sweat sodium concentration or chloride concentration accompanied by one or more of the following: the presence of chronic obstructive pulmonary disease, insufficiency of exocrine pancreatic function, meconium ileus, or a positive family history. The quantitative pilocarpine iontophoresis procedure for collection of sweat content must be utilized.

Two methods are acceptable: the "Procedure for the Quantitative Iontophoretic Sweat Test for Cystic Fibrosis" published by the Cystic Fibrosis Foundation and contained in, "A Test for Concentration of Electrolytes in Sweat in Cystic Fibrosis of the Pancreas Utilizing Pilocarpine Iontophoresis," Gibson, I.E., and Cooke, R.E., *Pediatrics*, Vol. 23:545, 1959; or the "Wescor Macroduct System." To establish the diagnosis of cystic fibrosis, the sweat sodium or chloride content must be analyzed quantitatively using an acceptable laboratory technique. Another diagnostic test is the "CF gene mutation analysis" for homozygosity of the cystic fibrosis gene.

The pulmonary manifestations of this disorder should be evaluated under 103.04. The nonpulmonary aspects of cystic fibrosis should be evaluated under the listings for digestive system (105.00) or growth impairments (100.00). Because cystic fibrosis may involve the respiratory and digestive body systems, as well as impact on a child's growth and development, the combined effects of this involvement must be considered in case adjudication.

Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. "Appropriate" means that the technique used is the proper

one to support the evaluation and diagnosis of the impairment.

E. *Bronchopulmonary dysplasia (BPD)*. Bronchopulmonary dysplasia is a form of chronic obstructive pulmonary disease that arises as a consequence of acute lung injury in the newborn period and treatment of hyaline membrane disease, meconium aspiration, neonatal pneumonia and apnea of prematurity. The diagnosis is established by the requirement for continuous or nocturnal supplemental oxygen for more than 30 days, in association with characteristic changes on medically acceptable imaging and clinical signs of respiratory dysfunction, including retractions, rales, wheezing, and tachypnea.

103.01 Category of Impairments, Respiratory System

103.02 *Chronic Pulmonary insufficiency*. With:

A. Chronic obstructive pulmonary disease due to any cause with the FEV₁ equal to or less than the value specified in Table I corresponding to the child's height without shoes. (In cases of marked spinal deformity, see 103.00B.);

Table I

Height without Shoes (centimeters)	Height without Shoes (inches)	FEV ₁ equal to or less than (L,BTPS)
119 or less	46 or less	0.65
120-129	47-50	0.75
130-139	51-54	0.95
140-149	55-58	1.15
150-159	59-62	1.35
160-164	63-64	1.45
165-169	65-66	1.55
170 or more . . .	67 or more . . .	1.65

or

B. Chronic restrictive ventilatory disease, due to any cause, with the FVC equal to or less than the value specified in Table II corresponding to the child's height without shoes. (In cases of marked spinal deformity, see 103.00B.);

Table II

Height without Shoes (centimeters)	Height without Shoes (inches)	FVC equal to or less than (L,BTPS)
119 or less	46 or less	0.65
120-129	47-50	0.85
130-139	51-54	1.05
140-149	55-58	1.25
150-159	59-62	1.45
160-164	63-64	1.65
165-169	65-66	1.75
170 or more . . .	67 or more . . .	2.05

or

C. Frequent need for:

1. Mechanical ventilation; or
2. Nocturnal supplemental oxygen as required by persistent or recurrent episodes of hypoxemia;

or

D. The presence of a tracheostomy in a child under 3 years of age;

or

E. Bronchopulmonary dysplasia characterized by two of the following;

1. Prolonged expirations; or
2. Intermittent wheezing or increased respiratory effort as evidenced by retractions, flaring and tachypnea; or
3. Hyperinflation and scarring on a chest radiograph or other appropriate imaging techniques; or
4. Bronchodilator or diuretic dependency; or
5. A frequent requirement for nocturnal supplemental oxygen; or
6. Weight disturbance with:

a. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall of 15 percentiles from established growth curve (on standard growth charts) which persists for 2 months or longer; or

b. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall to below the third percentile from established growth curve (on standard growth charts) which persists for 2 months or longer;

or

F. Two required hospital admissions (each longer than 24 hours) within a 6-month period for recurrent lower respiratory tract infections or acute respiratory distress associated with:

1. Chronic wheezing or chronic respiratory distress; or

2. Weight disturbance with;

a. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall of 15 percentiles from established growth curve (on standard growth charts) which persists for 2 months or longer; or

b. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall to below the third percentile from established growth curve (on standard growth charts) which persists for 2 months or longer;

or

G. Chronic hypoventilation (Pa CO₂ greater than 45 mm Hg) or chronic cor pulmonale as described under the appropriate criteria in 104.02;

or

H. Growth impairment as described under the criteria in 100.00.

103.03 Asthma. With:

A. FEV₁ equal to or less than the value specified in Table I of 103.02A:

or

B. Attacks (as defined in 3.00C), in spite of prescribed treatment and requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each inpatient hospitalization for longer than 24 hours for control of

asthma counts as two attacks, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of attacks.

or

C. Persistent low-grade wheezing between acute attacks or absence of extended symptom-free periods requiring daytime and nocturnal use of sympathomimetic bronchodilators with one of the following:

1. Persistent prolonged expiration with radiographic or other appropriate imaging techniques evidence of pulmonary hyperinflation or peribronchial disease; or
2. Short courses of corticosteroids that average more than 5 days per month for at least 3 months during a 12-month period;

or

D. Growth impairment as described under the criteria in 100.00.

103.04 Cystic fibrosis. With:

A. An FEV₁ equal to or less than the appropriate value specified in Table III corresponding to the child's height without shoes. (In cases of marked spinal deformity, see. 103.00B.);

or

B. For children in whom pulmonary function testing cannot be performed, the presence of two of the following:

1. History of dyspnea on exertion or accumulation of secretions as manifested by repetitive coughing or cyanosis; or
2. Persistent bilateral rales and rhonchi or substantial reduction of breath sounds related to mucous plugging of the trachea or bronchi; or
3. Appropriate medically acceptable imaging evidence of extensive disease, such as thickening of the proximal bronchial airways or persistence of bilateral peribronchial infiltrates;

or

C. Persistent pulmonary infection accompanied by superimposed, recurrent, symptomatic episodes of increased

bacterial infection occurring at least once every 6 months and requiring intravenous or nebulization antimicrobial treatment;

or

D. Episodes of bronchitis or pneumonia or hemoptysis (more than blood streaked sputum) or respiratory failure (documented according to 3.00C, requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each inpatient hospitalization for longer than 24 hours for treatment counts as two episodes, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of episodes;

or

E. Growth impairment as described under the criteria in 100.00.

Table III

(Applicable only for evaluation under 103.04A - cystic fibrosis)

Height without Shoes (centimeters)	Height without Shoes (inches)	FEV ₁ equal to or less than (L,BTPS)
119 or less	46 or less	0.75
120-129	47-50	0.85
130-139	51-54	1.05
140-149	55-58	1.35
150-159	59-62	1.55
160-164	63-64	1.85
165-169	65-66	2.05
170 or more	67 or more	2.25

103.05 Lung Transplant. Consider under a disability for 12 months following the date of surgery; thereafter, evaluate the residual impairment(s).

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