Breathing Problems Among Little People: When To Be Concerned

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Studies of the special problems with breathing that occur in little people have been ongoing for a number of years and at a number of medical centres. Among the most complex problems have been those in infants and children with achondroplasia, which have been the subject of specific study by the author, among others, through the cooperation of the membership of LPA, Inc. Studies in children with achondroplasia have enhanced the general understanding of the types of problems that can occur in other types of short stature as well. The following is a summary of our current understanding of breathing problems that are common among little people and are related to their different bone structure.

First, it is important to understand some facts about normal breathing function. When a person breathes, air enters the nose, and then goes to the hypopharynx (back of the throat) and then the larynx (voice box). Nasal breathing is automatic during sleep, but older babies and adults voluntarily breathe through the mouth when awake if the nose is blocked. Young babies have great trouble learning to do this. During sleep all the throat tissue becomes floppy, especially the tongue; mouth breathing is more difficult and maybe blocked by the tongue, which shows up as snoring. Air then enters the trachea (windpipe) and goes to the bronchi, which divide like tree branches and distribute the inspired air to the air sacs of the lung, where oxygen and carbon dioxide are exchanged. The movement of air occurs because of the downward movement of the diaphragm and bucket handle movement of the ribs, from the muscles between the ribs; these movements serve to enlarge the chest cavity. Impairment of diaphragm or rib movement decreases the amount of air moved in each breath. Breathing function is controlled by an important area of the brain known as the Respiratory Centre, which acts as a pacemaker for breathing. It is located near the beginning of the spinal cord, in an area of the brain called the medulla.

Understanding this allows us to understand some problems experienced by little people. Breathing problems in little people are of four major types: (1) problems with the bucket handle movements of the chest wall (2) decreased spaces for flow of air through the nose and hypopharynx (3) problems in the nervous system which affect the respiratory centre or muscle function and (4) individual problems of specific disorders other than the above.

Problems with the bucket handle movements due to abnormal bones cause Restrictive Lung Disease. People with this problem cannot get enough air in and out with each breath and they are unable to clear secretions and mucus from their lungs. Important symptoms include shortness of breath, rapid breathing and recurrent lung infections. This can occur with S-shaped spinal curvature (severe scoliosis), with severe shortening of the chest or with small chest cages, due to shortened ribs. Scoliosis occurs in many short stature conditions; breathing symptoms may worsen over time and are an important reason why

spinal surgery may be contemplated for these people. A short chest, related to a short spine, is an uncommon cause of Restrictive Lung Disease in older children and adults, but may be very serious or even fatal in certain infants. Small chest cages are characteristic of a number of short stature conditions; when extreme, this can be fatal, but when milder, may cause repeated problems with lung infection. Our studies in babies with achondroplasia showed that a small chest cage is frequent, but most affected babies outgrow this problem; some babies may need treatment until they grow. Treatment for Restrictive Lung Disease includes oxygen.

Problems with the air passages in the nose or hypopharynx cause Obstructive Upper Airway Disease. Obstructive Upper Airway Disease may be due to small bony air passages of the middle face ("dished out" face), crowded hypopharynx from a tiny jaw or thickened soft tissue in the breathing spaces, as occurs in storage diseases (mucopolysaccharldoses). The symptoms are inability to breathe through the nose, snoring and apnea (non- breathing) spells in all ages, and a multitude of symptoms in infants, including poor weight gain and inability to eat well. Mild cases may be merely an annoyance, but serious cases can lead to high blood pressure, heart failure or death. The condition may be made worse by the additional presence of Restrictive Lung Disease or neurological problems affecting the activity of the muscles in the breathing system. Our study of infants and children with achondroplasia found this to be a common problem, but few children were severely affected. It is present in a large number of older children and adults with achondroplasia and is made worse by being overweight. Sometimes, removing tonsils and adenoids helps, since it makes more space available in the throat. Other times, more drastic therapy is required. Treatment is dependent on the severity of symptoms. Obstructive Upper Airway Disease occurs in many short stature conditions and should be looked for anytime a person has the symptoms.

Breathing problems due to problems in the nervous system are less common, but may mimic other respiratory problems and therefore deserve mention here. Some short stature conditions may cause tightness or displacement of bones in the neck that may result in pressure, pulling or compression on the spinal cord and lower part of the brain (medulla). In achondroplasia, the opening of the bottom of the skull is very tight, adding to this possibility. This process may irritate or damage the respiratory centre and/or the nerves controlling the bucket handle motions of the ribs. Neurological symptoms such as weakness or abnormal coordination may or may not be present. Respiratory symptoms of compression of the medulla and spinal cord include weak breathing movements or apneic spells as well as a worsening of an otherwise mild case of Obstructive or Restrictive problems. The studies others and we have done about spinal cord compression in achondroplasia have shown that respiratory symptoms are common in babies and children with this problem. The same symptoms may occur in babies with other disorders who have compression. Treatment of these problems depends on making the diagnosis, which may be difficult, and relieving any compression which is present. Experience with a number of conditions in which compression occurs shows good results of surgery in general, but sometimes the problem, cannot be relieved if there has been sufficient damage to the nerves. Therefore, early diagnosis and treatment is of the highest priority for any baby with symptoms of apnea or unexplained breathing problem.

Specific breathing problems of other conditions include such problems as decreased lung function in people with storage disorders due to storage material in their lungs and problems with the larynx in people with diastrophic dysplasia. Finally it should be remembered that little people might get the same kinds of breathing problems as average statured individuals. Symptoms should always be evaluated carefully to diagnose the specific cause in any individual.

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