Thoracic Insufficiency Syndrome

Jonathan Fortunati- Biomedical & Electrical Engineering –University of Rhode Island

Out of the many healthy children born each year, there are an unfortunate few who experience complications due to various diseases. One such group of diseases is known as Thoracic Insufficiency Syndrome, or TIS. TIS is more of a term to describe a group of diseases that basically do the same thing: cause an inability of the thorax to support normal respiration or lung growth. This means, as the child grows, the growth of the lungs will be strongly hindered by the various diseases, causing pain, difficulty to breath, stunted growth, and in the most serious cases, death. There are three main types of diseases that make up TIS. The first is flail chest syndrome. This is when multiple ribs are broken, or not attached to the spinal column, causing a section of the chest wall to float around in the body. This causes intense labor while breathing because the floating chest wall works against the normal rhythm of breathing. This causes sever pain, and often, death. The second is sever scoliosis. Scoliosis is curving of the spinal column, like an s, which can shift the rib cage. This can cause sever humps in the rib cage and chest wall, limiting the growth of the lungs. The third is hypoplastic thorax syndrome, which consists of Jeunes syndrome, achondroplasia, jarchol evin syndrome, and ellis van creveld syndrome.

In general, TIS is due to a spinal or chest wall deformity, failure of the spine and rib cage to keep up with growth of the child. Disablement of the chest and spine function can cause the child to rely on nasal oxygen or ventilator support to breath. There are a couple of ways to solve the issues of TIS. One way is called spinal fusion. Spinal fusion can be used anywhere in the spine, but for TIS, it is used in the thoracic region. This technique fuses the two vertebra together, stabilizing the spine, and hopefully correcting an TIS complications. This can treat a variety of complications, mainly pain due to the diseases or neurological damage, like compression of nerves. The most relative disease treated by this method, is scoliosis.

There are two ways of performing the procedure ,allografting and autografting. These techniques are compounded with the bodies natural chemistry. Allografting is when cells from a donor of the same species, but not relatives, are transplanted into the human body. For example, taking bone marrow from one donor patient to another. Autografting is the same idea as allografting, but the cells are taken from another part of the patients body. For example, moving skin from the leg to the arm, for a skin graft. After the allografting or autograftign procedure, the body takeso ver. Osteoblats, cells responsible for bone formation, begin to create osteoids, which form the bone matrix. The bone matrix is then mineralized, forming a rigid bond between the two vertebrae. There are two types of spinal fusion. Inter body fusion is the process of spinal fusion where the intervertebral disk is completely removed between the two vertebrae. A graft is then placed, as well as a plastic or titanium disk, to allow the vertebrae to grow together. There is a fusion between the end plates, and this is known as 360 degree fusion. This method is sometimes accompanied by metal screws or pins. Posterolateral fusion is the second form, and is fusion between the transverse process. This method is usually accompanied by metal screws or pins. Generally, these methods can fix pain caused by certain types of diseases that would lead to a thoracic insufficiency syndrome, but when done in children (the target age group), it can prevent an already deformed and shortened spine from growing any longer. An undesirable side effect, but there is a product aimed at fixing this.

The VEPTR system is a solution to this problem. It stands for Vetrical Expandable Prosthetic Titanium Rib. The VEPTR runs, as it names states, vertically along the rib cage. Its main goal is to stabilize the ribs, to assist in TIS and allow the lungs of a child to grow normally. The device is implanted during an expansion thoracostomy operation. The device itself is unique, because it is not a rigid device, but a telescoping device that is attached to several ribs in the ribcage. The goal of the device is to act as a brace, keeping the rib cage and chest wall expanded. It also serves to help straighten the spine, correcting damage from scoliosis. The reason for the telescoping feature is so it can be adjusted as the child grows, lengthening it, and and continuously expanding the rib cage. The telescoping device is surgically adjusted every 4 to 6 months. This allows the patient comfort and normal breathing, which usually hindered by throacic insufficiency syndrome.

Being a relatively new procedure, there are on goign studies of its effectiveness and any complications due to the procedure. The first study done was a quality of life study. Being a more invasive procedure, a study was done to evaluate the quality of life of the patients who received the treatment. The study took 45 patients, diagnosed with TIS, between the ages of 5.6 and 10.8 years old. Before the procedure, they were given a health questionnaire for them to rate their quality of life, as well as their parents to rate their quality of life having to deal with the complication. Afterwards, once the VEPTR was implanted, they were given the survey again. The comparison of the surveys showed that the children's quality of life did not increase significantly, because they were still battling a disease. Although not a specific study, a website dedicated to parents who's children have undertaken this procedure, says otherwise. They report that their children are much happier, and live completely normal lives and activities such as sports are not limited at all by the implant.

A second study was done on children who received the implant and had Jeune syndrome or Jarcho Levin syndrome,that caused the TIS. 43 patients were taken out of a group of 214 who had the VEPTR device installed. Out of these 43, 4 children died of complications due to the disease. All four had jeune syndrome.. The goal of this study was to show that the device is not perfect, and cannot cure all patients. Because of this, surgeons should be aware of the possibility of post op death, and the likeness of death increases when the patient has Jeunes syndrome.

The device itself is covered by health care providers and in general costs between 40 and 70k. It is sometimes removed and sometimes not, depending on the condition and doctors preference. In extreme cases, the spine is also fused once skeletal maturity is reached.

Resources:

Mortality and life-threatening events after vertical expandable prosthetic titanium rib surgery in children with hypoplastic chest wall deformity.

Betz RR, Mulcahey MJ, Ramirez N, Flynn JM, Smith JT, St Hilaire T, Campbell RM.

J Pediatr Orthop. 2008 Dec;28(8):850-3.

Iatrogenic thoracic outlet syndrome secondary to vertical expandable prosthetic titanium rib expansion thoracoplasty: pathogenesis and strategies for prevention/treatment.

Nassr A, Larson AN, Crane B, Hammerberg KW, Sturm PF, Mardjetko SM.

J Pediatr Orthop. 2009 Jan-Feb;29(1):31-4.

http://www.veptr.com/

http://www.childrenshospital.org/clinicalservices/Site1171/mainpageS1171P4sublevel10Flevel12.html