

Multidisciplinary Chest Wall Deformity Clinic Initiated

by Kevin P. Moriarty, M.D., Consulting Staff Pediatric Surgeon

There are a variety of chest wall deformities that afflict children, the most common being pectus excavatum, pectus carinatum, and Poland's syndrome. Since the inception of the Chest Wall Deformity clinic at the Hospital in November of 1998, we have seen 51 patients in the clinic and performed 17 operations.

Pectus excavatum is characterized by the concavity (depression) of the sternum (breast bone) and occurs in approximately 1 out of every 400 children. This deformity often becomes more pronounced in early adolescence and can cause cardiopulmonary symptoms secondary to displacement. The most common presentation is one of psychological stress that children suffer as a result of this deformity. In the past, a lengthy invasive operation was required to correct the concavity of the sternum. Now the deformity can be corrected using a minimally invasive technique called the Nuss Procedure.

The Nuss Procedure places a bar under the sternum to remold the chest wall by applying an outward force to the sternum. In addition to the Nuss Procedure being minimally invasive, it also takes a fraction of the operating time. To date, 7 patients have had their pectus excavatum deformity corrected using the Nuss technique at Shriners Hospital.

Pectus carinatum is characterized by a forward buckling of the sternum and xyphoid. The deformity often becomes present in adolescence and worsens with growth. We have surgically corrected 5 patients with pectus carinatum. There is no minimally invasive procedure to correct this chest wall deformity. However, after consultation with the hospital's orthotics department, we have begun to treat this deformity without surgery by having the patient wear a custom molded vest (brace). By applying external pressure, this vest will attempt to remold the chest wall to a normal contour as with the Nuss Procedure.

Poland's syndrome is a rare syndrome that occurs in 1 out of 30,000 children and is characterized by a constellation of abnormalities, including syndactyly (webbing of the fingers), brachydactyly (short fingers), amastia (congenital absence of breast tissue) and absence of the chest wall musculature, as well as costochondral cartilages and ribs (cartilage between the ribs and breast bone). The patient may suffer from a variety of the components of this syndrome. The deformity can lead to a lung hernia, whereas, the only covering of the lungs is skin. Surgically placed prosthetic chest wall reconstruction is required in severe cases. Breast implants or pectoralis implants may also be placed by a plastic surgeon, as well as reconstructive surgery by a hand surgeon.

Kevin P. Moriarty, M.D. and Stanley Konefal, Jr., M.D., pediatric surgeons on the hospital's consulting



Pre and post operative correction of pectus excavatum using the Nuss Procedure.

Speech Pathologist Helps Children

Tcy Stannard, the mother of a former patient, recently joined the Shriners Hospital staff as its first Speech Pathologist. For 12 years, Ms. Stannard brought her son, Jud, to the neuromuscular clinic in the Hospital's Outpatient Department. Over the years, she witnessed many children with speech and language difficulties. As a result, she decided to change careers and return to school to become a speech and language pathologist. This newly created Speech Pathologist position fills a void for hundreds of patients with speech and hearing impairments. Ms. Stannard is responsible for evaluating patients, developing a communication plan and working with the family and school to implement the plan. Stannard is also an augmentative speech communication specialist, helping children to communicate by using assistive devices, ranging from communication boards to high-tech devices for non-verbal communication.

Her goals for the department are to include a lending library equipped with communication devices and switch activated toys that teach cause and effect. Another goal of the department is to teach children how to decrease excessive salivation and increase speech intelligibility by improving oral and facial muscle control with exercise.



Ms. Stannard is assessing 4 year old Maria for knowledge of cause and effect.

The Treatment of Juvenile Rheumatoid Arthritis with Borage Oil

Deborah Rothman, Ph.D., M.D., the hospital's Director of Pediatrics and Rheumatology recently completed a multi-center study which evaluated the treatment of juvenile rheumatoid arthritis with an oil extracted from the seeds of the borage plant. This oil contains gammalinolenic acid, a fatty acid that has anti-inflammatory properties. It has been shown to reduce joint swelling and pain in adults suffering from rheumatoid arthritis. This study was the first to investigate

whether it could be useful for children.

Twenty-eight children participated in a study for one year and were given either the borage oil or a placebo oil for a six-month period. Neither the families nor the physicians knew which oil the children took until the study was completed and the data analyzed. The results showed that the number of joints with active arthritis decreased significantly in the children taking the borage oil. (There were no concerning side effects.) This data was selected for presentation at the National Meeting of the American College of Rheumatology to be held in Boston in November.