

PROGRESS IN PEDIATRIC SURGERY – SELECTED ADVANCES IN LIFE SAVING PROCEDURES IN PEDIATRIC SURGERY

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Summary

Characteristic of pediatric surgery as a unique specialty dealing with the very special patient is presented. Main specific differences between surgical treatment in adult

patient and in child are discussed, pointing at the fact that pediatric surgeon takes care of a child starting as a fetus until achieving adulthood. Many if not majority children are treated surgically for various congenital malformations, which are never seen by surgeons treating adult patients. There have been many fields of pediatric surgery which have been developed during last 20 years and allowed for saving lives in many children. Author presents a selection of these fields and selection of the lifesaving procedures within those particular fields of pediatric surgery. There are presented comments on the progress in neonatal surgery, with particular underlining of fetal surgery as growing and still not fully explored field; progress in possibilities of correction of coagulation abnormalities and blood loss control during surgical procedures; and finally progress in liver transplantation in children which made this procedure an amazing and very successful procedure.

It is however important to say that the progress of pediatric surgery and final success in the treatment of each individual patient is based on progress of other disciplines and multidisciplinary approach and cooperation of many clinicians and other supporting staff.

1. Introduction

Pediatric surgery is not just a general surgery performed in smaller people. The smaller the child, the more differences in etiology of the diseases and indications for surgery, the more the differences in the physiology of the child and pathophysiology of diseases as well as the response of the child to the disease and surgical stress. The most prominent features of pediatric surgical patients are: many physiological mechanisms are not fully mature (immunological response, metabolic response, local and general response to infection); the child's body is in process of continuous growth, so it is connected with special needs in nutrition composition, caloric supply, vitamin and microelements; much smaller amounts of blood and other body fluids loss may lead to shock more readily than in adult; surgical techniques for children need to be much more precise and delicate than in adults; congenital malformations, often multiple, are the main indication for surgical treatment while most of these malformations are never seen by "adult" surgeon; treatment of these malformations must be based on very good knowledge of embryology and etiopathogenesis leading to these malformations during fetal development; surgical techniques should be aimed at reconstruction as much as possible of both anatomy and physiological function, bearing in mind that one operates on a growing organism.

These are only very general, and briefly presented differences between surgery in children and adults.

2. Scope of Pediatric Surgery

The scope of pediatric surgery may be expressed by the age range of pediatric patients. Nowadays our patients are not just children from birth till 14 or 16 years of age – but since 1981 pediatric surgery has become involved in fetal interventions and fetal surgery while at the other end pediatric surgery extends till 18 years of age when all growth processes are finished.

The scope of pediatric surgery may be also expressed as a spectrum of diseases or subspecialties within which children are treated surgically. Although several subspecialties

evolved from pediatric surgery (cardiac surgery, neurosurgery, orthopedics, etc.) this specialty is still very integral and not so much divided as in adults.

The part of pediatric surgery which differs most from adult type surgery is neonatal surgery. This is also the part of surgery which is most demanding for the surgeon, as well for as other cooperating specialists such as neonatologist and anesthesiologist. Assessing the level of the care provided by the pediatric surgical center can be effected by studying the results of surgical treatment of neonates with some so-called “index” malformations.

3. Progress in Pediatric Surgery as a Result of New Life Saving Procedures and Technologies

There is much new medical technology which has definitely influenced, directly or indirectly, the survival of patients treated by pediatric surgeons. It is impossible to describe them all but some should be high-lighted, such as new diagnostic or therapeutic techniques like new generations of USG, CT scans or NMRs, PET scans etc. These actually allow for three-dimensional reconstruction of examined regions of the body and provide extremely detailed visualization of any pathology prior to surgery. Further, there are such absolutely revolutionary life supporting systems as ECMO (Extracorporeal Membrane Oxygenation) and albumin dialysis of the liver - MARS system (Molecular Adsorbents Recirculating System), with which many children can now be saved that otherwise would die despite all possible conventional treatment. Additionally, many surgical devices and drugs aimed at minimizing or control of intra-operative bleeding (ultrasonic knife, argon beam, thermoablation, thermoresection, harmonic knife, fibrin glue, recombinant clotting factors and others) are now available that save life for patients with severe trauma, large malignant tumors or undergoing organ transplantation. The development of minimally invasive techniques and their miniaturization, has allowed endoscopic interventions in the fetus having severe life threatening malformations.

In this paper some of the advances which have influenced surgical practice in children will be presented. These include advances in fetal surgery, ECMO, MARS and new possibilities of hemostasis and bleeding control and surgery in children with liver failure, including liver transplantation.

4. Neonatal Surgery

4.1. Introduction

Surgical treatment of neonates with severe congenital malformations is in constant process of change based on better understanding of etiopathogenesis, embryological development and pathophysiology of congenital malformations and acquired diseases. Although surgical techniques themselves did not change during recent years, the changes in timing of diagnosis, including prenatal examinations, the timing of surgery, scope of procedures, preoperative and postoperative intensive care, influenced greatly and positively the outcome for newborns with many malformations. For example survival of neonates with congenital gastroschisis improved during last 20 years from about 50% to almost 100%. Similar improvement can be observed in such congenital malformations as esophageal atresia, omphalocele, congenital heart diseases and many other anomalies. There are,

however, malformations such as congenital diaphragmatic hernia, in which progress was not achieved to such an extent despite introduction of fetal interventions and new medical life supporting technologies - as for example extracorporeal membrane oxygenation (ECMO).

Simultaneous with the development of postnatal therapy during the last 25 years, a new surgical discipline is developing – surgery of the fetus.

4.2. Fetal Surgery

Fetal surgery originated with Michael Harrison, who performed in 1981 the first open surgical procedure in the fetus with urinary obstruction – vesicostomy. Since then several new procedures have been introduced, some of them abandoned later as ineffective or unsafe for the fetus or mother.

In 1981 for the first time a urinary catheter was introduced into the urinary bladder of the fetus under ultrasonographic guidance (so called vesico-amniotic shunt), instead of open fetal surgery for urinary outlet obstruction (Harrison). Within a few following years the first successful resection of fetal lung mass (cystic adenomatoid malformation of the lung) was performed (Harrison, 1984), followed by fetal surgery for congenital diaphragmatic hernia (Harrison, 1989). In subsequent years the first resection of sacrococcygeal tumor in the fetus and repair of spina bifida were done.

Since these pioneering experiences, much knowledge was gained and most procedures were reevaluated or changed in many congenital anomalies and new procedures were introduced. But fetal surgery is still only performed in a very limited number of centers.

4.2.1. Types of Fetal Surgery

There are several types of fetal surgery from less invasive (amniotic sac puncture) to most invasive for both mother and the fetus (open fetal surgery).

4.3. FIGS Procedures

Most commonly used are so called FIGS procedures (Fetal Image Guided Surgery) performed as diagnostic or therapeutic procedures. These procedures are: cordocentesis (fetal blood sampling), amniocentesis, amnioinfusion and amnioreduction or shunt procedures. The shunting made by FIGS procedures are: vesico-amniotic shunts (for decompression of congenital obstructive uropathy), pleuro-amniotic shunts (for decompression of congenital hydrothorax) or ventriculo-amniotic (for decompression of congenital hydrocephalus).

Enthusiasm and indications for shunting procedures have rather diminished during the last few years, as they do not really improve prognosis for the patient as well as treated organs. This statement is particularly important for fetuses with central nervous system malformations, renal dysplasia and unilateral obstructive uropathy. Vesico-amniotic shunts still have important role in the treatment of progressive renal failure in the fetus caused by subvesical obstruction of the urinary tract.

4.4. FETENDO Procedures

The development of laparoscopic instruments which were also developed for operations in newborns, resulted in their application for fetal surgery. Procedures utilize the minimally-invasive techniques of video-surgery in the fetus, are via laparoscopy in the mother. During FETENDO procedure a balloon is introduced into the trachea in the fetus with pre-natally diagnosed congenital diaphragmatic hernia with severe lung hypoplasia. Other procedures include disruption of posterior urethral valves in male fetuses with urinary tract obstruction. It has also been used for amniotic bands division and cord division.

Of particular interest is the use of the FETENDO technique in fetuses with congenital diaphragmatic hernia. It replaced open fetal surgery during which the diaphragmatic defect was closed. Open procedures were followed by a high number of complications, and did not change much the course after delivery, as pulmonary hypoplasia was not improved. Open surgery was replaced by the, experimentally checked, idea of closure of the fetal trachea in fetuses with diaphragmatic hernia, thus causing distension and growing of hypoplastic lungs due to constant production of the fluid within the fetal respiratory tract. This idea was combined with FETENDO techniques by which a detachable balloon is placed into the fetal trachea endoscopically between 22-28 weeks of gestation, and then removed by the same technique about 1-2 weeks before delivery. This procedure is reserved for fetuses with most severe lung hypoplasia, confirmed by measurement of so called lung-head ratio (LHR). FETENDO should be performed only in cases when LHR is below 1.0. The real advantage of the FETENDO procedure for diaphragmatic hernia is still under investigation.

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Biographical Sketch

Prof. Piotr Kalicinski MD, PhD, born 1955, graduated from Warsaw Medical Academy, is specialist in pediatric surgery and clinical transplantation and surgery. His main professional and scientific interests are: liver and biliary surgery, hepatic tumors, dysganglionoses, congenital malformations and neonatal surgery, vascular surgery and transplantation. He is one of the pioneers of pediatric kidney and liver transplantation in Poland. He has established the first program of liver transplantation in children in Eastern Europe in 1990 and first living related liver transplantation program in 1999. Actually he is the Chief of Transplant Program and Head of Department of Pediatric Surgery and Organ Transplantation, Childrens Memorial Health Institute in Warsaw. His professional carrier includes training in: Pittsburgh University, UCLA at Los Angeles, King's College Hospital in London, Necker Hospital in Paris and Medical School of Hannover, Germany, Kyoto University, Japan.

Prof. P. Kaliciński is a member of European Union of Pediatric Surgeons Associations, International Pediatric Transplant Association (councilor), member of Steering Committee of PLUTO (Pediatric Liver Unresectable Tumor Observatory). He is also a member of editorial boards of: *Pediatric Transplantation*, *European Journal of Pediatric Surgery*, *Annals of Transplantation*,

Prof. Piotr Kalicinski is one of the founders of Polish Organization for Organ Transplantation Coordination – POLTRANSPLANT. He is President of Polish National Transplant Committee, advisory committee by the Polish Government.

He published over 250 papers and chapters, more than 300 abstracts.

Prof. Piotr Kaliciński was awarded by Polish Ministry of Health, Polish National Scientific Committee, Polish Transplant Association, World Academy of Medicine and Polish Academy of Success.