جراحی کودکان در ایران و چالش های پیش روی آن

دکتر هوشنگ پورنگ

Pediatric surgery in Iran and challenges

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1. What is subspecialty in pediatric surgery? is It different from adult surgery
2. Establishment of pediatric surgery discipline in Iran
3. Who are Pioneers of pediatric surgery in Iran?
4. Where is your position as a pediatric surgeon in the Iran's medicine now?
5. How many pediatric surgeons are there in Iran?
6. How many pediatric surgeons do we need in Iran?
7. How much we have been successful in obtaining our purposes in pediatric surgery?
8. Where we should be going in pediatric surgery?
9. Which are our future problems?
10. Are there in which part our problem?
11. Which group has Overlap working with the field of Pediatric Surgery?
12. Who can better to do the operation and management of an infant or a child with these complicated anomalies?
13. What is the mission of a pediatric surgeon?
14. What is the responsibility of ministry of health, treatment and Medical Education in the promotion and improvement of this subspecialty in Iran? Have they done their responsibility well?
15. With lack of interest of young general surgeon to pediatric surgery, what would happen to children with complicated congenital malformations that need surgical treatment?
16. Do our country need pediatric surgeons more or other branches of surgery?

تاثیر شکاف کام درتکامل اولیه تکلم

دکتر فاطمه درخشنده

Effect of Cleft Palate on Early Speech-Language Development

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The literature suggests that prior to 3 years of age; toddlers with cleft palate demonstrate limited vocabulary, restricted sound inventories, and the emergence of compensatory articulation errors. Babies with cleft palate are at a distinct disadvantage during early vocal development. Throughout the prelinguistic period at least, the baby is obligated to engage in vocal practice without the normal division between the oral and nasal cavities provided by the hard and soft palate and, in many cases, the absence of normal articulatory contacts in the anterior portion of the hard palate. These anatomical constraints have the potential to influence the baby's vocalizations in several ways: -producing sounds that do not require linguapalatal contacts. -avoid production of early stop consonants during babbling. Conductive hearing loss may distort the auditory signal and impair vocalizations. The majority of children with cleft palate will require the services of a speech-language pathologist (SLP) at some point in their lives. In such cases the SLP may not provide direct services to the baby but will instead serve as a resource for parents, who need information about the effects of a cleft on speech and language development. SLPs may meet with parents and their baby for the first time just before or immediately after the palatal repair. This is unfortunate because valuable time has been lost during a critical period in communication development. Ideally, the SLP on a cleft palate team or early intervention team should initially meet with parents when the infant is no more than 3 months old.

کاربرد فیبرین طبیعی و مواد مصنوعی باهم درترمیم فیستول بعد از ترمیم شکاف کام

دکتر مرجان جودی، دکتر مهران هیرادفر، دکتر رضا شجاعیان، دکتر رضا نظرزاده

Combination of Native Fibrin Adhesive and Prosthetic Materials in Post-Palatopasty Palatal Fistula

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Aim: Palatal fistulas represent a challenging problem for surgeons caring for patients with cleft palate. The incidence of fistula occurrence after palatoplasty is ranged 11%–34%.all conventional attempts in fistula closure show disappointing results. Fistula repair with combined prosthetic and native materials are rarely discussed so we presented our results in this filed.

Methods: Patients with palatal fistula after palatoplasty were admitted and blood sampling and processing was performed to obtain native fibrin glue. Limited palatal flaps were created around the fistula edge circumferentially and a Prolene mesh a little bigger than the fistula was placed under the flaps to cover the fistula completely and fixed with 5-0 Vicryl sutures. Fistula edge was refreshed and the prosthesis was covered with native fibrin adhesive and a free bucal mucosal graft was laced over the fistula and fixed with 6-0 Vicryl sutures.

Results: among five patients with palatal fistula reconstruction with prosthesis and fibrin glue was planed meanly in 4 month after the first operation. Mean age was 2.7 years. Patients were fallowed for at least 2 months and successful results were observed in four cases. Recurrent fistula was seen in only one case while the fistula decreased in size significantly from 1.3mm2 to 0.6mm2 and the mesh was preserved in place without any infection or other complications. A limited debridement and redo fibrin glue coverage of fistula was performed and the fistula was obliterating after the second operation.

Conclusion: We suggest this technique as a safe, effective, and simple method of palatal fistula repair with significantly lower fistula recurrence rates but further experiences on more cases need for better judgment.

نقش فلپ های فارنژیال خلفی در درمان اختلالات ولوفارنژیال

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The Role of Posterior Pharyngeal flap in the Treatment of VeloPharyngeal Dysfunction

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Background: The velopharyngeal apparatus is the combination of soft palate & pharyngeal structures that regulate airflow from the lungs & larynx through the mouth for oral sounds & through the nose for nasal sounds. Throst – cardamone uses Velopharyngeal inadequacy as the generic term for any type of abnormal velopharyngeal function and divides that in to 3 categories by etiology: Velopharyngeal (VP) insufficiency, VP incompetence & VP mislearning. VP dysfunction is used as the generic term for any type of abnormal Vp function (D, antonioz). The most common cause of VPD seen by plastic surgeons is clefting of the secondary palate. Structural causes of VPD due to cleft palate occur in 1 in 2000 live births and VPD occurs in approximately 20% of children after palatoplasty. The most common symptom in VPD is hyper-nasality. In VPD a careful approach should be used to localize the cause, if the problem is a VP gap then is surgical intervention can be selected. The pharyngeal flap is the preferred surgical approach in patients with good lateral Pharyngeal wall motion who have a persistent central gap resulting from poor Palatal motion as is common following repair of a cleft palate. one large series of 500 patients with pharyngeal flap repair, reported normalization or resolution of hyper-nasality in 90% of patients.

Method: This article is a report of the results of posterior pharyngeal flap in the treatment of VPD in 8 patients after cleft palate repair. Nasoendoscopy and oudiometric and speech assessment exams were used to evaluate the patients before and after surgical intervention.

Results: There was a significant improvement in nasoendoscopic and oudiometric and speech assessment exams among our patients after pharyngeal flap repair.

Conclusion: Pharyngeal flap repair suggested as an effective method in treatment of velopharyngeal dysfunction.

ترمیم شکاف لب و کام در بیمارستان کودکان فرانسوی کابل- افغانستان

دکتر محمد طارق رحیمی

Cleft lip and Palate at FMIC

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Generally cleft lip (CL) and palate (CP) is the defect on the upper lip and the roof of the mouth and form embryological it's result of internal nasal, external nasal and maxillary buds fusions anomaly. Second common malformation after club foot, affecting approx one in every 1000 births worldwide. This malformations is more in Asian than the other races, incidence of CL is 2/1 according to male /female and CP 2/1 female/male. Diagnosis made by ultrasound exam during pregnancy or simply by physical exam after births.

Clinically it has four types Cleft lip ,Alveolar Cleft ,Hard Palate cleft and Soft Palate cleft .The only option for treatment is surgery ;closure of lip and palate which has different type and modifications and operation timing.

In our Institution FMIC the best and accepted timing for CL is three months age and for CP six months and according of our experience and finding the best procedure for CL closure for functional and cosmetic side is Tennisson, Simple Z, Double Z, Malek, Skoog and modification by Onizuka, for CP closure is Veau-kilner –Wardill technique, Bardach Method-Two Flap technique and Sommerlad procedure. After six months usually we perform cleft lip and palate repair is some time in one stage, for CP repair up to five years age we do muscle closure by sommerlad technique and after that age repair of palate and Pharyngoplasty. For secondary Lip surgery we do by Onizuka technique (modification of Millard 3), we're trying to operated very carefully and not just simple lip closure and adhesion but real Cheilo-septo-rhinoplasty. According to our study lip adhesion distorting the normal lips muscles and tissues

Since 2007 up to now (Dec.2011) we had operated 271 cases of CL and CP, in which 177 CL and 94 CP, Male and Female ratio 174/97 totally, for CL 103/74 and CP 44/50.According to age about 132 patients up to 6 months, 59 patients up to one year, 32 patients over 10 years and others over them, In 25 cases CL and CP repaired in one stage, two cases of CP with Teacher Collins's syndrome.

لاترال ویدئو فلوروسکپی در تشخیص نارسائی ولوفارنژیال (گزارش جراحیهای ثانویه شکاف کام در کودکان)

دکتر فاطمه درخشنده

Lateral Videoflouroscopy in Diagnosis of Velopharyngeal Insufficiency (The report of Palatal Secondary Surgeries in Children with cleft palate)

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Objective: To describe the use of Lateral Videoflouroscopy in palate re-repair in the treatment of velopharyngeal incompetence (VPI) and analyze the results in a consecutive series of patients operated on by a single surgeon. Design: Prospective data collection, with perceptual and instrumental (lateral videofluoroscopy) assessment of speech and velar function. Patients: seven consecutive patients with previously repaired cleft palates and symptomatic velopharyngeal incompetence (VPI) and evidence of anterior insertion of the levator veli palatini underwent palate re-repairs by a single surgeon (Brian Sommerlad) from 2008 to 2011 in Isfahan Cleft Clinic. Interventions: Palate re-repairs, with radical dissection and retropositioning of the velar muscles, were performed using the operating microscope. Main Outcome Measures: Pre- and postoperative perceptual speech assessments using the universal parameters for reporting Speech outcomes in individuals with cleft palate (Farsi version), measurement of velar function on lateral videofluoroscopy.

Results: There were significant improvements in hyper-nasality, nasal emission, and nasal turbulence and measures of velar function on lateral video-fluoroscopy, with improvement in the closure ratio, velopharyngeal gap. Conclusions: Video-fluoroscopy is a radiographic procedure for visualization of all aspects of the velopharyngeal portal during speech, through the use of several standard views. Palate re-repair has been shown to be effective in treating VPI following cleft palate repair in patients who have had a previous attempt at muscle dissection and retro-positioning. Palate re-repair has a lower morbidity and is more physiological than a pharyngoplasty or pharyngeal flap.

فتق دیافراگم تشخیص وپی آمد قبل از تولد

Congenital diaphragmatic hernia: Antenatal diagnosis and outcome

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Introduction: Congenital diaphragmatic hernia (CDH) is a potentially correctable anatomical defect that continues to represent a significant cause of neonatal death. The incidence of CDH is between 1 in 2000 and 1 in 4000 live births and it accounts for about 8% of all major congenital abnormalities. It can be detected during fetal life when screening ultrasonography demonstrates herniation of the intestine and/or the liver into the thorax.Clinical problems are due to associated abnormalities and the neonatal risk of severe respiratory failure induced by pulmonary hypoplasia and persistent pulmonary hypertension.

Aims: We want to describe antenatal detection and outcome of CDH in neonates.

Methods: A retrospective study of 19200 live births during4years between Jan .2008 and Dec2011was performed and **5** cases of CDH were detected after birth.

Results: We reviewed the medical records of 5caces of CDH both neonates and their mothers , all mothers had prenatal careand4of them had at least two ultrasound examinations of normal fetus during pregnancy .4 neonates were born with cesarean section and one with normal vaginal delivery,**5**min APGAR score was >**7**in3 cases. 4 neonates were expired within 6 hours after birth and one was alive and was sent to a referral hospital for further management.

Conclusion: Studies show increasing antenatal detection rate of CDH about 54% and increasing survival rate about 50-80%, whenever antenatal diagnosis is made, it is advisable to direct the mother to a tertiary perinatal center in which all the necessary obstetric, neonatal and surgical skills are concentrated.

نخستین گزارش موارد ترمیم استنوز هیپر تروفیک پیلور به روش لاپاروسکوپیک در ایران

دکتر مهران هیرادفر، دکتر احمد بذرافشان، دکتر مرجان جودی، دکتر رضا شجاعیان، دکتر رضا نظرزاده

The First Experience of Laparoscopic Pyloromyotomy for Hypertrophic Pyloric Stenosis in Infant from IRAN

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Introduction: Conventional Ramstedt pyloromyotomy was an accepted method in treatment of IHPS for decades. In 1991 the first successful endoscopic pyloromyotomy was performed in the world and recently the single port endoscopic repair is introduced in this filed. We started endoscopic treatment of IHPS for the first time in Iran (Mashhad University of Medical sciences) From the May of2010.In this article we report the results of this modality in IHPS.

Method and patients: From the may 2010 to February 2011, 11 cases were included in this study. Diagnosis was at the base of Ultrasonography. Laparoscopy performed by inserting the supraumblical 3-5 mm port for camera and 1-2, 2-3 mm stab wound for instruments. Intra-abdominal Pressure was maintained in the range of 3-6 mmHg. Superficial electro-cautery followed by cold knife incision and dissecting the hypertrophic segment by grasping forceps was performed in all cases. Feeding started the day after operation.

Results: 11 cases were included in study (male- 3 female).In all cases we finished the procedure by endoscopy. There was no need for conversion. In 9 cases feeding was started at the day after operation and was tolerated well except in 2 cases that tolerated later. In one case omentum protruded through the 3^{mm} stab wound site which was excised and returning to abdominal cavity under sedation. No any other complication occurred during hospital stay.

Conclusion: MIS in management of IHPS is an acceptable method with a good result and at least with a better cosmetic outcome.

تظاهرات حاد و تاخیری فتق دیافراگم در کودکان گزارش یک مورد از بیمارستان مفید

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Late and Acute presentation of congenital diaphragmatic hernia in Mofid Children's Hospital

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Aim: Congenital cystic lesions of lungs are uncommon, but have similar embryologic and clinical characteristics. Some of these lesions have late and acute presentation with unusual clinical manifestation.

Case report: 7 months old girl that presented to pediatric emergency department with respiratory distress and mild cyanosis. On physical examination child was tachypenic and restless with decreased breathing sound in left thorax. In chest X-ray there was huge pneumothorax in left thorax with shifting of mediastinum. Patient was admitted immediately to PICU for respiratory support. Surgical consult was done. Resident surgeon asked the pediatric surgeon for chest tube insertion. Evaluation of patient by pediatric surgeon showed her respiratory distress is mild and her O2 saturation was 93% and her vital signs are stable. Chest tube insertion cancelled and patient kept in PICU under close observation for more investigation. Chest CT was done in the next day that confirmed cystic lesion of left lung. Left thoracotomy was done that showed a single huge lobulated cystic lesion from lingula lobe of left lung that inflate with inspiration and compressed the normal lung. Resection of cyst was done. The left lung expanded well. Patient has had uneventful recovery and discharged from hospital in a few days.

Conclusion: In patient under 1 year old, cystic lesions were discovered by respiratory distress; and in patients over 1 year old signs of infection were the most important clinical features. Early recognition of these relatively rare congenital cystic lung lesions would lead to the immediate, proper surgical intervention.

دکتر زینب ال بلوشی، دکتر احمد نجیب، دکتر یوسف ال شخصی، دکتر یاسر والی

Laparoscopic splenectomy in Sickle Cell Disease children

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Background: Sickle cell disease (SCD) is the most common heamoglobinopathy in the Arabian Gulf region and in Oman. Though the natural history of SCD in the Caribbean and African haplotypes is that of autosplenectomy by age of 5 years, the Arab –Indian haplotypes children maintain their spleen enlarged till older age group.

They are at risk of splenic sequestration, hypersplenism and massive splenomegaly. Over the past 15 years splenectomy was practiced in Sultan Qaboos University Hospital (SQUH) through an open approach. Recently from December 2009 we are shifted to laparoscopic approach. We report our experience of open versus laparoscopic splenectomy in children, age 3-14 years old with SCD from July 2008 to December 2011.

Subjects and methods: All children with SCD who had splenectomy over the past 3 years in SQUH were included in this study.

They were divided into 2 groups. Group A (n=22) those who had open splenectomy. Group B (n=24) those who underwent laparoscopic splenectomy.

Results: The mean ages for both groups were 7.2 years. The indications for splenectomy were recurrent splenic sequestration (93.5 %) and hypersplenism (6.5 %).The mean size of the spleen in both groups were 13.7 cm. Post surgical pain severity in Group A (5.9) and in B (3.5) with p value <0.0001. Complications related to SCD such as acute chest syndrome was 18.2% in group A versus 8.5% in group B. There were no vaso-occlusive crises or Hemorrhage in both groups. Post-operative pyrexia was 36.4 % in first group versus 25 % in second group with p value of 0.05, Restoration of normal appetite and bowel habits were almost equal in both groups. Post op blood transfusion (3 patients in group A as well 3 patients in group B. Mean Length of stay (8.8 days in Group A versus 6.2 days in group B with p value of 0.0001Risks of recurrence due to missed splenosis are not studied yet in both groups.

Conclusion: Laparoscopic splenectomy is much safer, feasible and with much less post surgical pain, complications and Hospital stay in SCD patients.

تجربه ترمیم ساده با گره خارج بدنی فتق قدامی دیافراگم با لاپاروسکپی درشیرخواران

دکتر محمود سعیدا، دکتر احمد خالق نژاد طبری

The Experience with a Simplified Technique of Laparoscopic Anterior DiaphragmaticHernia Repair in Infants Using Extracorporeal Knotting

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Background & Purpose: Although laparoscopic repair of anterior diaphragmatic hernia in older children has been increasingly accepted as the method of choice, there are still some concerns and even controversies about routine use of it in infants because of their small working space. We report our experience with the simplified endoscopic repair of this type of hernias in infants with only one or two working-ports using extracorporeal knotting. Patients and Methods: Since Aug 2009 to Feb 2011,three infants with anterior diaphragmatic hernia operated on laparoscopically using 5-mm 30° telescope inserted by open technique via umblicus and one (or two according to severity of adhesions and access difficulties) working-port in the both sides of umblicus and after meticulous reduction of the herniated viscera (transverse colon in two and left liver lobe in one), the para-sternal diaphragmatic defect (two left-sided and one bilateral), repaired with 3-4 separate 2/0 silk stitches. The needle introduced directly into the abdomen over the hernia and getting a bite of the posterior rim of the defect with a U-stitch, the needle brought-out thru the abdominal wall at the same entrance point, and after tying extracorporeally, each knot buried in the subcutaneous fat layer.
Results: Three infants (a 4-months 5-kg, a 7-months 6-kg, and an 8-months 8-kg male babies), who presented with persistent respiratory symptoms and had documented retrosternal hernias on chest X-ray, underwent laparoscopic repair. The mean operative time was 40 minutes without any conversion. We had no intra-operative complication and there was no problem with CO2 pneumoperitoneum. All patients discharged on the third postoperative day and followed by control X-ray at 1, 3 and 6 months. Our first patient had normal control CXR at 3-months postoperatively, but his 6-mo postoperative X-ray showed a small asymptomatic recurrence. At laparotomy we found a small neglected right-sided parasternal defect, along with the healed previously repaired left-sided defect.
Conclusion: Considering the small working space and to overcome the difficulties of intracorporeal knotting in small infants, we recommend this simplified laparoscopic technique using extracorporeal knotting for all parasternal hernias even in infants and small children as the routine method. We also recommend meticulous checking the other side to rule out bilaterality.

تعیین ریسک فاکتورها در فتق دیافراگم مادرزادی

دکتر سید جواد نصیری، دکتر مریم قوامی عادل

Determination of risk factors which affect the Short term outcome in newborn with isolated congenital diaphragmatic hernia in an educational children hospital

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Introduction: Despite a lot of progress in medical and surgical care, mortality of posterolateral congenital diaphragmatic hernia (CDH) is steel high (40 – 60%). Various factors affect this high rate of mortality. In this study it tried to define the factors which affect on the short term outcome of these neonates.

Method and material: We performed a retrospective cohort study in which all patients with diagnosis of isolated congenital diaphragmatic hernia were included. Neonate with associated cardiac or other anomalies were excluded. The variables which were evaluated were as follow: demographic data, time of presentation(onset), first PaO2, first PaCO2, presence of Persistent Pulmonary Hypertension (PPHN), highest PaO2, liver position, stomach position, Gestational age, laterality and outcome(discharge alive or dead).The data were analyzed by SPSS 16.0. **Results:** 52 patients were included, with57.7% male. Fifty nine percent of the patients were discharged alive, 40.4% were dead, in 75% of the patients, left side and in 1.9% bilateral sides were involved. The variables which were significantly affected outcome were as below; PPHN, Time of presentation, first PaO2, first PaCo2. Gestational age, laterality, liver and stomach position have no significant affect on short outcome of the patients. The highest PaO2, in the first or second day of management by conventional ventilation (before operation), were in the range of 90.6 – 235, in> 60% of the patients who afterward were expired

Discussion: In newborns who born alive with CDH the mortality rate (despite applying many kind of modern facilities) is still high. Various contributing factors which affect the prognosis of the disease have been suggested. In our study, PPHN, the first PaCO2, the first PaO2 and time of presentation, were the most important factors which was significantly related to the outcome. The other variables were not

Conclusion: 1- Newborns who deliver alive with isolated CDH, presentation of symptoms in the first 6 hours of life, with the first PaCO2 > 50, PaO2<60 and with symptoms of PPHN, would have very bad prognosis. 2- Since in >60% of the patients who expired, have shown highest PO2 > 90.6 (honey moon period), the authors suggest that it could be an alarm that we should look for a different type of ventilation. 3- Could ECMO substitute this kind of ventilation? We hesitate.

نخستین گزارش موارد ترمیم هرنی دیافراگماتیک مادرزادی به روش توراکوسکوپیک در ایران

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The first experience of Thoracoscopic congenital diaphragmatic hernia repair in neonate from Iran

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Introduction: Minimally invasive surgery (MIS) for treatment of CDH is introduced recently. The advantages for this method include less pain, minimal tissue injury, and better cosmetic result. For the first time in Iran we started the Thoracoscopic repair of CDH from April 2011 in Mashhad university of Medical Sciences.

Patients and Methods: 13 cases were operated by Thoracoscopic method in Dr Sheikh pediatric Hospital from April 2011.Under the general anesthesia, in lateral position, a 5^{mm} port was inserted for camera. 2 other 3^{mm} stab wounds were used for instrument insertion. Intra-thoracic pressure was 6^{mmHg}. In 6 cases primary repair was done by 3-0 Prolene material. In 5 cases we use the thoracic wall for primary repair and in 2 cases prosthetic material was used. Chest tube was inserted in all cases.

Result: Median age was 5 days (Range 3 days to 7 months). Duration of operating time was 135±27.8 min. the defect was in the left side in 12 cases. In one case due to preoperative iatrogenic trauma to stomach due to chest tube insertion, thoracic cavity was contaminated but successful thoracoscopic repair of stomach performed. In one case due to insufficient space of abdominal cavity laparoscopic iatrogenic eventration created before the Thoracoscopic approach. Early in-hospital recurrence occurred in one case (7.7%). This case managed by open prosthetic repair. In a mean of 181 days follow up, any recurrence, mortality or morbidity occurred.

Conclusion: This is the first report of Thoracoscopic repair of CDH in neonate from IRAN. It seems that Thoracoscopic repair of congenital diaphragmatic hernia is feasible with accepted result although the cost is more compare to conventional methods.

تجربه ۱۰ ساله ترمیم فتق دیافراگم (از سال ۱۳۸۰ لغایت ۱۳۹۰) در بیمارستان کودکان بهرامی

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A ten years experience of congenital diaphragmatic hernia repair in neonates from Bahrami children's Hospital (2001- 2011)

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Introduction: and aims: congenital diapharagmatic hernia (C.D.H) is a common congenital anomaly that occurs with diaphragmatic defect in embryonic period and interning of abdominal organ in the thorax cavity. Incidence CDH is one in 2000-5000 live births and most common clinical presentation is respiratory distress.

Neonatal group with CDH that present within 6first hours usually they have a high mortality. In past believed that emergency portative intervention has a better survival, But today most believed that delayed operation after stabilization of the patients is have better survival Today Stabilization of the patients became by strategy of to use of mechanical ventilation and ECMO without barotraumas to the lungs.

Methods: in this cross sectional study medical data of all admitted neonates, infants and older children in Bahrami children's hospital related to Tehran University Medical Sciences and operated for CDH from 1380 to 1390 entered to this study and by SPSS version analyzed

Results: Total patients were 116 neonates with (55.2%) 64 male and (44.8%) 52 female.

86(74.1%) of them had delivered by cesarean section and 30 patients by normal delivery.22 (19%) patients had near related family and 6cases (5.2%) late related family and88 (75.8%) cases had not related family

8(6.9%) patients had prenatal diagnosis and 60 cases had post natal diagnosis .36 (31%) were infants and 12 patients were more than 2years old . Most clinical presentation at admission time was respiratory distress in 90(77.6%), 18(15.5%) cases had GI problems and remained 8 (6.9%) cases has other problems. pre operation the patients intubated by tracheal tube. Of 116 operated patients only 16 cases died.

Conclusion: In this study there was a significant etiological relationship of mortality with preoperative history of intubation .diagnosis time, weight at surgery .so diagnosis at neonatal period (p. v=0.02%), weight<3kg (p. v=0.0006) and preoperative duration and tracheal intubation (p. v=0.002) and mortality significantly was high

Prenatal Diagnosis and Fetal Intervention in Fetus with High Airway Pressure, Stenosis, Obstruction and Lung Anomalies

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Introduction: Some giant neck mass like teratoma and high airway obstruction syndrome (CHAOS) and CCAM and agenesis of lung, heart and kidney if delayed to detect and some induce hydrops which are fatal for fetus diagnosis early and fetal intervention will cure the fetus.

Method: Data collected from articles and discussed the detail for the colleague

Results: Fetal giant neck masses such as teratoma can grow to such large proportions that the fetal airway becomes distorted and obstructed with cervical teratoma; the mass effect pulls the lung into the apex of the chest and result pulmonary hypoplasia. In addition to obstructing the airway these masses also can compress the esophagus resulting in polyhydramnios which can lead to uterine irritability and preterm labor.

Congenital high airway obstruction syndrome (CHAOS) results large echogenic lungs flattened or inverted diaphragm, dilated airway distal to the obstruction and fetal ascitis and/or hydrops. Some of these patients with hydrops tolerated progressive hydrops 12 weeks in utero but some led to fetal demise and some delivered by exit procedure.

Congenital cystic adenomatoid malformation (CCAM) with hydrops in utero resection of the enlarged pulmonary lobe reduced mediastinal shift and allowed expansion of normal lung tissue. 3 types of CCAM are detected. Type 1 50% large cystic lesion few in number (1 to 4) very favorable. Type 2 40%, 1 to 5 small cyst is more commonly associated with other congenital anomalies. Like cardiac, skeletal, genitourinary, hydrocephalus and CDH. Type 3 10%, these are usually micro cyst lesions that cause shifting of the organs normally found in the chest, these mean that tracheal heart and lung are shifted and compress for this reason heart function may be less than normal this can lead to hydrops.

In largemonocyst fetal intervention with Thoraco-Amniotic shunt must be done and in hydrops Fetal intervention CCAM resection led to resolution of the hydrops impressive in utero lung growth and neonatal survived.

Conclusion: The prenatal diagnosis obstruction airway early prevent hydrops and

The Exit procedure when diagnose late potentially salvage lethal condition.

نتائج ترمیم آترزی مری در بیمارستان علی اصغر ۱۳۸۶ - ۱۳۸۹

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Results of Esophageal Atresia repair in Aliasghar Pediatric Hospital 2007- 2010

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Introduction & Purpose- The most life-threatening anomalies of the esophagus are the variants of esophageal atresia and tracheoesophageal fistula which associate with other anomalies. The aims of this review are to analyze results of esophageal atresia in our center.

Material & Method: Between "1386 to 1389 "the record files of all neonates with esophageal atresia and TEF admitted in NICU of Aliasghar Hospital were evaluated. The demographic data, associated anomalies, types of TEF, surgical complications, survival and causes of mortality evaluated.

Results- 62 neonates were investigated.31 (50%) patients were female and 31 (50%) were male with mean gestational age 36.5 weeks. Themean weight of patients was 2.5 kilograms. The most common type was type C (87.1) and then type a (11.3%).The most common associated anomaly was cardiac anomaly (40.3%).There was no complication after operation in 77.4% of patients. Anastomotic leakage occurred in 6.5% of patients. The most common post operative complication was stricture (9.7% -6patients) that 5 patients respond to dilatation and 1 underwent surgery. In long term follow up 6.45% had GER that 5% need fundiplication. Mortality rate was 17.7% which sever congenital heart disease was the main cause of early mortality.

Conclusion- There were small differences between our findings and the literature. Only mortality in premature infants was more than the references. Then our postoperative and NICU care must be improved.

نکات برجسته ترمیم موفق آترزی مری بدون سوند مری در۳۸ مورد

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Surgical Highlights in Successful Tubeless Repair of EA/TEF Experience with 38 cases

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Introductions and Purpose: A well-founded concern exists among many practitioners over anastomotic leakage in early postoperative period after an EA/TEF repair. Recent studies demonstrate that the undue stress and discomfort, which is associated with chest drain or NG tube, cause restlessness in patients and adversely affect the healing process. A tubeless repair, where no chest drain or NG Tube is used after EA/TEF repair and some management maneuvers, obviates this unnecessary suffering and thus eliminates the risk of leakage.

Material and Method: The records of 38 infants who underwent EA/TEF repair between 2003 and 2009 were analyzed. Demographics, associated malformations, birth weight and postoperative complications were recorded. The procedure involved posterior extraplural thoracotomy to leave the azygus vein unsevered, division of TEF and end-to-end anastomosis of pouches as Haight;s model with 6-0 vycril sutures. We did not use chest drain, NG Tube or feeding tube. All patients underwent primary repair and received TPN postoperatively for 5 days.

Results: During a 6-year study period, a total of 38 cases of esophageal atresia with tracheaesophageal fistula including 21 male and 17 female infants were treated. Seven of these patients were diagnosed in the prenatal period by a third trimester ultrasound. Of the 38 cases, 23 (60.5 %) had associated anomalies including cardiovascular (13), ARM (6), GU (5), GI (4) and V (3) . Eighteen patients had a birth weight between 2000-2500 g and the remaining twenty were between 2500-3500 g. In majority of the cases the diagnosis was confirmed at birth by inability to pass an orogastric tube. In most of the cases the gap between two esophageal pouches was about 3-4 cm. Postoperative barium swallows did not reveal any leakage and the early postoperative days of hospitalization were uneventful. The oral feeding began at the fifth postoperative day after the esophagography confirmed no anastomotic leakage. **Conclusion:** There is no need to use any chest drain and or NG tube routinely for protective mechanism after EA/TEF repair. To achieve successful repair in these conditions some technical points and management maneuvers must be considered.

گزارش سه دهه درمان آترزی مری در بیمارستان کودکان مفید

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Three decades report of management of ESOPHAGEAL ATRESIA in Mofid Children's Hospital

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Introduction & Aim: It has been more than 30 years since the reconstruction of esophageal atresia (EA) was performed in the Mofid Children's Hospital (MCH). We reviewed the historical changes in management and treatment results of patients with esophageal atresia that admitted in MCH for three decades according to existing data of 4 studies.

Method: We collected the data of 4 studies that have been done as thesis 2 for MD degree and one for specialty degree and one for subspecialty degree and analyzed the data of 283 consecutive patients treated for esophageal atresia in our center from1360 to 1389 that divided to 3 periods, 1360-1369, 1370-1380, and 1381-1390.

Results: 283neonates with EA admitted in MCH.1360-69, 23 cases, 1370-80,129 cases and 1381-90, 131 cases. .Admission age was between1day to12day old.1360-69, 60.8% of neonates were male, 1370-80, 49% and 1381-90, 48%were male. 1360-69, 56.5%of neonates had birth weight more than2500 gr.1370-80, 63% and 1381-90, 58%.In1360-69, Most common type of this anomaly was type C(EA with distal fistula) 60.8%, type A(without fistula) 17.3%,typeB (proximal fistula) 4.3%,typeD (distal and proximal fistula) 8.6%and H type 0%. 1370-1380: type C 85%, type A 5%, type D 1.5%, H type 2% and congenital esophageal stenosis 2%. 1381-1390: type C 86.6%, type A 3%, type D 2% and H type 1.5%.1360-69: In prenatal ultrasound polyhydroamnious reported in17.3%, 1370-80:35.2%and1381-1390: 90.4%.1360-69: 56.5%of neonates had associated anomalies. Most common associated anomaly was cardiovascular 26.1%, anorectal13%, genitourinary3%,skeletal6% and other13%.1370-1380: 47.35%had associated anomalies,cardiovascular18.45%, gastrointestinal11%, skeletal11.7% , genitourinary3%,right side descending Aorta0.8% and VACTERL associated anomalies 6.5%. 1381.1390:59% of neonates had associated anomalies, cardiovascular21.9%,anorectal9.45%,genitourinary7%,skeletal7.25%, gastrointestinal 9%, right side descending aorta 3%, and VACTERL anomalies 7%.1360-1369: 61%of esophageal atresia underwent primary repair of which 38%had gastrostomy.20% underwent gastrostomy and esophagostomy and the others with long gap EA underwent fistula ligation, gastrostomy and esophagostomy.1370-1380: 81.2%performed primary repair of which 22.3% had gastrostomy, the others underwent staged repair with gastrostomy.1381-1390:immediate primary repair performed in 96%. 1360-69: overall mortality rate was 83 %(9% before and74%after surgery).1370-80: 37%. (11%before and26%after surgery).1381-90: 24.5 %(3.5%befor and 21% after surgery).Increase of survival is due to NICU improvement in MCH.1360-69:post-operative complications included:8.6%anastomotic stricture,4.3%anastomotic leak,4.3%gastroesophageal reflux,4.3% recurrent tracheoesphageal fistula,8.6%wound infection ,and4.3%gastrostomy leakage.1370-80: 29.3%%anastomotic stricture,10.6%anastomotic leakage,4.9% recurrent tracheoesphageal fistula, gastroesophageal reflux didn't determine correctly. 1381-1390: 32%anastomotic stricture,3%anastomotic leak,55%gastroesophageal reflux,2.3% recurrent tracheoesphageal fistula, and 6%gastrostomy leakage.After1381 the rate of anastomotic stricture increased perhaps duo to increase in primary repair in long gap atresia.

Conclusion: the patients who are treated nowadays for esophageal atresia in MCH are diagnosed earlier, with better supportive care and improved surgical techniques. Therefore, the major defect of our center is the lack of careful and long term surveillance of these patients.

نتایج درمان آترزی مری در بیمارستان کودکان فرانسوی کابل – افغانستان

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Esophageal Atresia

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Introduction and Aims: Incidence of esophageal atresia (EA) is estimated between one each 2500 till 5000 live birth based on literature. We present the result of different operative technique which is used for treatment of EA since 2006 in FMIC.

Methods: All patients with EA which are included in this study were registered and treated in pediatric surgery department of FMIC since 2006. 10% of patients were coming directly to the hospital and 90% were referred from other centers.

Results: Eighty five new cases of EA were recorded since 2006 in pediatric surgery department of FMIC.

Incidence of EA is 2.5/10000 of live birth, according to the literature. The sex ratio was 1.55 (M/F) .The mean birth weight was 2250+_ 750 g and gestational age was 36+_4.00. Prenatal diagnosis was suspected only in 1% of cases. According to the anatomical classification, they were 6 patients with type I, 1 patient with H type and 78 patients with type III. Diagnosis was made within the first 24 hours in 20%

Of cases and surgery was planned before 48th hours in 90% of cases of type III. 6 patients died before surgery because of sever septic condition, 15 patients died before discharged from hospital. Associated

Malformations were present in 55% of cases.

Length of stay was significantly longer in the group premature and low birth weight and the group patients which were transferred from other centers.

Conclusion: This is the first study which discussed EA in Incidence, sex ratio, Gestational age, anatomical classification, associated anomalies, operative techniques, complications and survival rate in pediatric surgery department of FMIC, Kabul/Afghanistan.

بررسی ارزش تشخیصی باریوم انما در هیرشپرونگ نوزادان

دکتر محمد هادی رفیعی

Evaluation of diagnostic value of barium enema in Neonatal Hirschsprung's disease

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Background: Hirschsprung's disease (HD) is a congenital defect in pediatric patients, which is manifested by bowel obstruction in neonates or constipation in children. The gold standard of diagnosis is absence of ganglionic cells in myenteric neural plexus and submucosa of bowel. With regard to barium enema is a simple, less invasive and more available method than rectal biopsy, diagnostic evaluation of barium enema compared with rectal biopsy as a diagnostic method in HD is important.

Methods: In a cross - sectional study, 21 children with clinical manifestations of HD that referred to Al-Zahra hospital of Isfahan , Iran were assessed from 2005 through 2009.The method sampling was census. Barium enema and rectal biopsy were carried out for all cases. Data after collected entered to computer and analyzed by SPSS soft ware. The Ch- square and T student tests were used for data analysis. Also Sensitivity, specificity, positive predictive value and negative predictive value of barium enema were calculated for identifying of the HD.

Results: From 21 studied children, 9 (41.9%) were boy. The mean age of the children was 15 ± 10 days. The sensitivity, specificity, positive predictive value and negative predictive value of barium enema for diagnosis of the HD were 50%, 100%, 100%, and 60%, respectively. Diagnostic value of barium enema was calculated according to groups ($p<0.05$). Mentioned above, that; there were significant differences among them

Conclusion: The results of the study showed that barium enema cant applying for HD diagnosis. Therefore, it is recommended considering all four aspects of clinical manifestations, barium enema, anorectal manometry and rectal biopsy in diagnosis of HD. The gold standard for diagnosis of HD is rectal biopsy.

(تجربه ده ساله) آترزی مری بررسی ریسک فاکتورها درمورتالیته

دکتر حیدر علی داوری، دکتر مهرداد حسین پور

Mortality in Esophageal Atresia: Evaluation of risk factors (Ten years experience)

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Background: Esophageal atresia (EA) is a common congenital anomaly and survival in these patients still is not so good. In this study, we evaluated the mortality and its risk factors in patients born with esophageal atresia.

Methods: A database of 206 consecutive patients treated for EA was developed in ST-Zahra hospital of Isfahan between 1994 and 2004

Results: In this survey 206 patients with esophageal atresia were studied (127 boys, 79 girls). The most common type of EA was type C (86.4%). Total mortality rate was 36%. Mortality rate was more common in patients with associated anomalies (43.7% vs. 29.9%) P=0.004 and in patients with preoperative pneumonitis (36.4% vs. 32.7%) P=NS. It also was more common in low birth weight and preterm babies (p<0.001). Patients who had operation during the first two days of birth, had low mortality in 21.3% vs. 31.5% P=0.003. Mortality was more common in patients who had gastrostomy and secondary repair in comparing with primary repair (35.5% vs. 20.2%) P=0.02. The most of the patients died during neonate period (84.1%) and the most common cause of death was sepsis. Mortality rate was 43.9% in the first five years of the survey and 27.1% in second five years (P).

Conclusions: Despite significant improvement in our patients' survival in recent years, but it was higher than worlds'results. For more improvement of EA survival, we should improve operation techniques, supportive care and ICU management. It also important to diagnose this anomaly and also associated anomalies and operate these patients as soon as possible

دیلاتاسیون تنگی مری بعد از عمل آترزی مری

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Dilatation of postoperative Esophageal stenosis in Mofid Children's Hospital

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Background: Esophageal atresia (EA) is a common anomaly in neonates. Esophageal anastomotic stricture (EAS) is one of the most frequent complications of EA repair and is usually treated with esophageal anastomotic dilatation (EAD).

Materials and methods: From Jan 2005 to June 2011, 165 neonates with EA were treated in our institute. Primary anastomosis was done in 163 cases. In 20 cases, esophageal mechanical dilatation was done after presentation of clinical symptoms of EAS. In 20 cases as prophylactic early dilatation (ED) group based on operative judgment for EAS developing, early dilatation was performed two weeks after EA repair. The incidence of EAS, number of dilatations, first time of dilatation and complications were recorded in two groups. The efficacy of dilatation was evaluated by clinical examination and esophageal contrast study. All patients received medical treatment for gastroesophageal reflux (GER).

Results: Six cases with isolated EA and 10cases of mortality due to severe associated anomalies and sepsis were excluded. In 40 of 163 neonates there was operative judgment for EAS formation (Long gap between both ends, tension on anastomsis, proximal esophageal myotomy and the ischemia of distal segment) as prophylactic ED group underwent 1 to 3 dilatation (average 1.5). Two of 20 did not response to dilatation and needed revision of anastomosis.

The mean follow up period was 12 months.

Conclusion: EAS following EA repair is related with several factors. Early EAD is an effective method of treatment with high success rate in patient suspected to develop the EAS.

نقش تیم درمانی در بیماران آترزی مری در دو بیمارستان کودکان دانشگاه علوم پزشکی تهران

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Role of Team Management on Esophageal Atresia and TEF in 2 Children's Hospitals of Tehran University of Medical Sciences

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Introduction and Aims: Esophagealatresia/Tracheo-esophageal fistula is common congenital anomalies, by advanced in perinatology and NICU in recent years its mortality and morbidly rates were reduced but still is high, Aims was to assay the outcome of these patients in our hospitals in recent 5 years

Method: medical files of 43 patients underwent surgery for EA/TEF from 2004 to2009 in 2 children hospitals of TUMS retrospectively reviewed demographic, associated anomalies, types of atresia, operative, mortality, morbidity data accumulated and analyzed, incomplete files excluded from the study

Results: records43 patients 23male (53.5%); mean BW=2620gr (Birth Weight: 16<2500gr&27>2500gr) respectively reviewed, There were ; Drooling in 39 patients (90.6%), unable feeding 40(93%),,weight loss13(30.2%).preoperative pneumonia 10(23,3%) . associated anomalies 17(39.5%) CHD (most VSD,PDA).Most patients (86%) had C type atresia .mean age at Operation was 8.26 days(Ranged 2-22 days) .a premature neonate with pneumonia died before operation. 5 cases underwent staged operation and remainder primary repair ,Post-operative death was :4(9.4%),2 due to sepsis,2 had staged operation (one prematurity & CHD , another down syndrome and CHD).From 4 deaths, 2 was female &2 male. Relationship between death and sex statically was not significant (P=1.000).Post-operative complication were anastomosis Leakage: 16 (37.2%), recurrent fistula 3(7%), GERD: 23(53.5%).Correlation between complication and mortality and sex was not significant. Relationship of length of gap and CHD with complication were not significant (P=0.345), but relation of BW<2500grand overall complication was significant (P=0.042)

Conclusion: with reducing of the mortality rate in recent years , still associated sever CHD and respiratory infection and prematurity and LBW chromosomal abnormalities are the most causes of death and increased survival dictated a requirement of long follow up and cooperative team working

نخستین گزارش موارد ترمیم آترزی مری به روش توراکوسکوپیک در ایران

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The First Iranian Group Report of Thoracoscopic Esophageal Repair

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Introduction: Conventional Thoracotomy for repair of EA-TEF is a well known method for decades. The first successful thoracoscopic repair was done in 2000 in the world. Until 2004 the number of successful thoracoscopic repair remain minimal in the world .Up to 2010 less than 300 cases were reported in the world. The first successful repair of Thorcoscopic EA-TEF in Iran was done in Pediatric Dr sheikh hospital (Sarvar Fundation) (Mashhad University of Medical Sciences) in Sept 2011. Until yet 6 successful cases were repaired By MIS. In this article we report our experience about the MIS surgery in EA-TEF.

Methods and Cases: From February 2010 to April 2012, 16 Cases of EA-TEF were eligible for MIS. Excluding criteria were weight under 2 Kg and significant cardiac anomaly. In the first 9 cases we couldn't finish the operation and converted them to conventional method. In remaining 7 cases, except in one we able to finish all of procedure thoracoscopic. The position was 45 degree semi- prone position. We use 4 port (One for camera, 2 for grasping and one for holding the lung).TEF managed by exposure repair and cutting it Esophageal repair was performed by 5-O Vicryl suturing. CT inserted in all case.

Results: Mortality was zero. In 6 successful repair (3 male, 3 female), the average length of time was 220 minute. One well controlled leak occurred in this group that was managed by conservative methods. Stricture occurred in 2 cases that were successfully managed by dilatation.

Conclusion: It seems that EA repair by MIS is a possible way. It is much more difficult in compare to CDH and other endoscopic repair in neonatal field and need more experience. Some advantages are: cosmetic and prevents chest wall deformity and wing scapula. On the other hand Permissive hypercarbia, hypoxemia and increase the cost are negative points in this method. Further data can finally guide us to clearly choose the best way for treatment of EA cases.

نتایج ترمیم آترزی مری در بیمارستان کودکان امیرکلا ۱۳۷۳ - ۱۳۸۶

دکتر سهیل اوصیا

Evaluation of Esophageal Atresia Repair in Amirkola Children's Hospital from 1994 to 2007

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Introduction: The most prevalent congenital anomaly of esophagus is atresia with or without trachoesophageal fistula, occurring in about 1 in 2500 to 3000 live births. There was associated anomaly in these patients that causes mortality in these patients.

Aims: The aim of this study was to evaluate the esophageal atresia at Amirkola Children's Hospital.

Method: This study was performed on 37 patients with atresia that were corrected surgically in Amirkola Children's Hospital during 1994 to 2007.Dataincluded age, gender, birth weight, type of atresia associated anomaly, post surgical complication, duration of hospitalization, hospital mortality, time of ventilator and kind of surgery were recorded for all patients.

Results: In this study there were 23 male and 14 females. The mean gestational age was 37.2±1.7 weeks. The mean birth weight was 2601±505 gr. Type of atresia was esophageal atresia with distal trachoesophageal fistula in 75.6% and assassinating anomaly in 37.8%.Post surgery complication and mortality was 59.5% and 29.7% respectively. Time of hospitalization was 16.5±11.7 days. Time of ventilator was 5.5±3.1 days and mean age in surgery was 3±2.3 days. Kind of surgery were thoracotomy with closure the fistula and esophageal anastomosis in 70.3% and gastrostomy with esophagestomy in 24.3% and surgery throw the neck with closure of fistula in H type of TEF in 5.4%.

Conclusion: The most prevalent type of atresia is c-type and its morbidity and mortality decreased over time despite the equality of surgery type due to development of NICU care

فریبا نوایی(کارشناس پرستاری)، دکتر هدایت الله نحوی

Esophageal Atresia and TEF: NURSIG CARE and Parents Recommendations

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Introduction: Esophageal atresia (EA) is a condition in which the esophagus does not form completely in a developing baby. Tracheoesophageal fistula (TEF) is a condition in which an abnormal channel (fistula) connects the trachea to the esophagus.

There are several types of tracheoesophageal fistula and esophageal atresia. In the most common type (About 85% of children), the upper part of the esophagus has a closed end and the lower part of the esophagus connects to the trachea. About 1 in 4,000 children are born each year with TEF, EA or both. About one-third of these children are born early.

Symptoms of TEF and EA: The most common signs of TEF/EA are: Breathing problems:

Coughing or choking Babies who have TEF only usually have mild coughing or respiratory symptoms when they are feeding. Babies with TEF/EA may have problems that are not directly caused by the conditions, but are linked to them. Doctors call this the VACTERL association. Each letter stands for a possible problem that is associated with TEF/EA:

Diagnosis: before your baby is born based on the results of a prenatal ultrasound.

After your baby is born, doctors diagnose TEF/EA by trying to insert a nasogastric tube (NG tube). The tube passes through the baby's nose, down the esophagus and into the stomach. In children who have EA, the NG tube hits a blocked end, usually in the mid- chest. Doctors can see this on an X-ray. Usually they need no other diagnostic test. To find TEF without EA, doctors sometimes use special tests: Barium swallow test. The barium helps the digestive tract show up on X-rays.

Surgery is the only treatment for TEF and EA. Most babies with the conditions will have surgery soon after they are born. Some babies who have other, associated problems may need to wait a short time before having surgery.

Gastrostomy: The surgeon makes an opening through the wall of your baby's belly and places a tube into the stomach. This provides a safe way to give your baby nutrition until they can feed by mouth. It also lets doctors give your baby medicine, if needed. The surgery team will teach you how to take care of the tube and the skin around it, and how to feed your child through the gastrostomy the time babies need to stay in the hospital after surgery for TEF and EA varies. If your baby is healthy and the repair was simple, they may need to stay for a minimum of two weeks. If your baby has other health problems, an esophageal atresia with no fistula, or if the repair was delayed, your baby may need to be in the hospital for weeks to months.

Gastro esophageal reflux: The most common long-term problem for babies with TEF/EA is a condition called gastroesophageal reflux: We usually can treat this with medicine. As they grow older, children with TEF/EA usually can eat normally. They probably need to eat more slowly and chew more thoroughly than people who did not have TEF/EA

Outcome of Treatment in 51 Patients with Fecal Incontinency in Three Hospital of Tehran

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Background: Fecal incontinence is a problematic issue in all societies. We can divide patients with fecal incontinence into two groups according to surgical management. Group one has normal and intact perineal body and anal sphincter but abnormal innervations. Patients with myelomeningocele, cerebrospinal trauma, and mental retardation are in this group. In the other group the perineal and anal sphincter have normal innervations but the muscles are not intact. Patients with defect in anal sphincter or mislocation of the sphincter after anorectoplasty or trauma are in this group. We have to evaluate these patients with EMG, MRI, radiography, endoanal ultrasonography, muscle stimulator and providing sphincter mapping (especially in group two) for proper diagnosis and management.

The purpose of this study is to evaluate the long term results of surgical management and complications in patients with fecal incontinence that referred to the author.

Method: It is a retrospective descriptive study on 51 patients with fecal incontinence in three hospitals in Tehran between May 1993 and February 2011.

Results: patients with abnormal pelvic innervations and normal anatomy underwent ACE (Antegrade Colonic Enema) procedure and were dry with 2-3 times a week enema through the appendicostomy. There were few complications like stomal stricture (in 3 patients) and prolapse (in 2 patients) that were corrected surgically. In 27 patients from 37 patients in group 2 with perineal and anal sphincter problems, sphincter muscle defect and anorectal displacement after anorctoplasty for ARM were the causes of fecal incontinence, six patients had abdominal pull-through and the other 21 had postero sagitol anorectoplasty (PSARP). The other causes of fecal incontinence were: anterior anus (3 patients), megarectosigmoid (2 patients), rectal prolapsed (2 patients) sever anal stenosis (1 patient), perineal trauma (1 patient) and cloacal anomaly (1patient). After careful evaluation with Barium Enema, EMG, MRI, radiography, endoanal ultrasonography, muscle stimulator and anal sphincter mapping, according to the results, different surgical procedures such as re-do anorectoplasty, anal sphincter repair, anorectal reposition, megarectosigmoid resection performed. In partial rectal mislocation the most of defects were at 6 and 12 o'clock in lithotomy position. After the corrective operations 27 patients had complete continence. Nine patients had soiling (1-2 times a week) and in one of the patients because of non efficient anal sphincter ACE procedure performed.

Conclusion: Most of the pediatric fecal incontinences are controllable. As sphincter defects after anorectoplasty were in 6 and 12 o'clock specific attention to this issue require during primary anorectoplasty in ARM.

گزارش نحوه درمان دو مورد آنومالی های پیچیده آنورکتال و ژنیتال در کودکان

Report of Two Complicated Anorectal and Genital Malformation: Total Mobilization of Pelvic Organ and Prinea to Midline

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Complex anorectal malformations (CARM) are group of unusual heterogonous defect.

In this type of defect, surgeon may encounter bizarre cases with complex anatomy we had two cases of female complicated anorectal and genital malformations that underwent total mobilization of all structures of perinea to midline.

Case I: 6 month old female with skin covered bladder, pubic diastasis, ectopic anus, vagina, urethra to the right side of median cleft and lipoma on the left.

VCUG showed bladder in right pelvic with low capacity and trabeculation with no reflux.

Ultrasound and DMSA showed left kidney agenesis. Endo-sonography and pelvic MRI showed normal internal sphincter and weak external sphincter on right side.

Muscle stimulator around ectopic tight displaced anus was acceptable but had no muscle in median cleft and left side. Closure of pubic diastasis with iliac osteotomy and double barrel sigmoid colostomy was done at 8 months age. In second operation, right displaced anus, vagina and urethra and all muscle complexes underwent total mobilization to median position.

Case II: 8 Month old female with complicated anorectal malformation that underwent double barrel colostomy on day 8 of birth.

On examination she had hemivagina, urethra and anus at the right side of median cleft and other hemivagina at the left and lipoma between two vaginas.

Ultrasound and explorative laparatomy showed left side pelvic kidney and bicorn uterus and duplicated vagina.

In the second operation, median cleft lipoma excised and total mobilization of anus, urethra, RT hemivagina and muscle complex to the midline was done and the two hemivagina sutured together

In the third operation the wall between two hemivaginas divided by ligature bipolar.

Conclusion: Complicated ARM is correctable but each of them represents a peculiar challenge and will require a great deal of ingenuity and imagination in order to reconstruct them.

بررسی فاصله دیستال رکتوم تا پوست در بیماران باآنورکتال مالفورماسیون نوع high با لوپوگرام

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Determination of Distance between Distal Rectal Margin and Skin in High Type Anorectal Malformation by Distal Loopogram

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Purpose: This study was performed to compare ordinary distal lopogram and controlled augmented pressure distal lopogram in accurate diagnosis of recto urinary fistula. In high imperforated anus with divided sigmoid colostomy.

Methods: This was a clinical trial study that conducted in Alzahra Hospital in 2010. In this study infants with imperforated anus (n=12) who undergo surgery and prior operation, first, undertaken ordinary colostogram and then, underwent pressure monitoring distal lopogram and the outcomes of both technique were compared.

Results: At first of study ordinary distal lopogram was done for all of 12 patients that rectourinary fistula was detected in one patients (8.3%) but according to pressure monitoring distal lopogram 10 cases (83.3%) of rectourinary fistula were detected and there was significant correlation between detection of recto urinary fistula and use of pressure monitoring distal lopogram (P value =0.004).

Conclusion: The pressure monitoring distal lopogram with 250-360 mmHg of controlled pressure has more effectiveness than ordinary technique in diagnosis of recto urinary fistula.

مقایسه ترمیم یک یا چند مرحله ای شیر خواران آنوس بسته با فیستول رکتو وستیبولار

Comparison of Single and Multi Stage Reconstruction Procedure for Infants with Imperforate Anus and recto-vestibular fistula

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Abstract: The most common defect is rectovestibular fistula. This malformation may be repaired during the neonatal period without a protective colostomy. These operations UN fortunately represent the most common source of complications in such patients.

Methods and Materials: The decision on repair this malformation primarily or to open a colostomy should be based on the experience of the particular surgery who is going to perform this procedures.

Within years 1996 to 2008, fifty six infants with imperforated anus of recto-vestibular fistula

Have undergone single or multistage reconstruction procedures in our hospital in this retrospective prospective study, method of treatment, outcomes and surgical complications in these patients were evaluated. Fourteen infants (group A) have been within 1-13 days of birth (mean= 4 days) and 42 infants (group B) within 3-5 months (mean=3.5 months). Infants of group A underwent multi stage procedure including colostomy + anorectoplasty + colostomy closure. Patients in group B received intermittent dilatation of recto-vestibular fistula and after 3 months of age single stage anorectoplasty was done. Early and late surgical complications were considered for recording.

Results: As seen in table daily follow up of patients in groups A and B consisted of the following: posterior vaginal wall detachment, superficial wound infection, deep wound infection, dehiscence, rectal retraction, vaginal retraction, fistula recur, fecal incontinence, anal stenosis, rectal mucosal prolapsed, constipation, enteric adhesions and surgical wound scar.

Conclusion: Decision making for selecting single or multi stage procedures for reconstructing imperforated anus mainly depends on surgeon's experience. Comparing with group and in group B, perineal superficial wound infection was more prevalent, but no scar tissue was observed and hospital stay was less.

Neonatal Primary PSARP in High Imperforate Anus without Colostomy

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Background: The standard approach to management of high imperforate anus is colostomy in the newborn period followed by posterior sagital anorectoplasty (PSARP) at 6 to 12 months old.

The purpose of this study was to determine whether a non-stage repair by primary PSARP in the new born period cloud be performed without clear detriment to the patient's functional result. Methods: Between 1993 TILL 2003 totally 30 newborns with high imperforate anus who underwent primary PSARP without colostomy were studied retrospectively 17 males who had. Rectourethral fistula (15 bulbar and 2 prostatic type) and 13 females (10 rectovestibular and 3 rectovaginal fistula) All patients had ano-sphinctric evaluation after 3 years old, and early postoperative complications too. All cases were studied by sonography, echocardiography, lower vertebra X-RAY and finally routine blood and urine laboratory tests. Incontinence was defined as fecal soiling at least twice a day, and patients requiring more than office dilatation were described to have stricture formation.

Result: All patients recovered well and have been followed for periods ranging from 1.5 to 10 years. There were 3 cases of postoperative wound infection, but no anastomotic dehiscence Stricture, formation and fistula recurrence were seen. In cases of constipation (6) and fecal incontinence (5) there were associated anomalies such as sacral dysplasia, kidney dysplasia, VDS and VUR. There were no complications in other cases.

Conclusion: the most important factor is patient selection, and one-stage PSARP spares the patient the morbidity of additional surgeries in the standard multistage approach for high imperforate anus.
These preliminary results suggest that one-stage posterior sagital anorectoplasty is a safe and suitable approach to the management of high imperforate anus without clear determine to future bowel function.

دکتر حمید فروتن

Fetal Surgery: The indications, procedures and outcome

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Advances in prenatal diagnosis of the fetal anomalies now permit the intrauterine intervention. The rate of anomalies is high but many are treatable if diagnosed and treated prenatally.

Meningomyelocele is one of the most common anomalies that can be treated at 22-24 weeks of gestation with excellent results. Most of these patients, more than 70% will have normal lower extremities with urinary and fecal continence.

Patients with congenital diaphragmatic hernia can get benefit from intrauterine intervention and temporary blocking of trachea.

The limbs of patients with amniotic constrictive bands can be salvaged if treated antenatally.

Extrauterine Intrapartum treatment (EXIT) can be life saving for huge neck masses and large CCAMs.

Intrauterine interventions are also effective in patients with posterior urethral valve and UPJ obstructions.

Twin-twin transfusion syndrome (TTTS) can also be treated with fetoscopy and laser intervention.

Still rate of abortion with fetal surgery is about 20-30%. New advances, minimally invasive therapy, experience through animal study can improve the survival and the results.

Good team work, including perinatologists, fetomaternal surgeons, Pediatric surgeons, Pediatric neurosurgeons and urologists, genetists, radialogists and anesthesiologists and good nursing care are necessary for fetal surgery intervention.

کیست هیداتیک در کودکان، یک مطاله در تبریز

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دکتر حبیب عبدالهی

Hydatid Cyst in Children: a study in Tabriz, Iran

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Background: Hydatid disease is usually caused by the cestode Echinococcus Granulosus for which humans are an intermediate host. It occurs throughout the world and is especially common in sheep- and cattle-raising regions such as Middle East including Iran. Although the liver is the most frequently involved organ in adults, the most common organ of involvement in children are lungs.

Methods: A total of 21 patients of hydatid cyst disease who were admitted to the Tabriz Children's hospital, Iran have been studied in the present study. The diagnosis was made on the clinical suspicion followed by the ultrasound examinations of the liver and chest computed tomography. Surgery was chosen as the main treatment followed by medical therapy and tight control radiologic examinations.

Results: The mean age of the patients was 8.5 ± 3.6 years of age. Fifteen patients (71.4%) were male and 6 patients (28.6%) were female. The involved organ was liver in 5 patients (23.8%); while it was lung in 12 patients (57.1%) and both liver and lung in 4 patients (19.0%). In the patients with liver hydatid cyst right lobe was involved. In patients with lung hydatid disease, 2 patients (12.5%) had the cyst in left lung, 12 patients (75.0%) had the cyst in right lung and 2 patients (12.5%) had the cyst in both lungs.

Conclusion: Hydatid cyst is a potential infectious and surgical concern in Iran and knowledge of its clinical presentation, diagnosis and treatment is of clinical importance. The results of this study reveal that hydatid disease should be considered as a differential diagnosis in children with liver and lung masses. In addition, unlike adults, lung hydatid disease should be kept in mind rather than liver hydatid disease in children.

STERNAL CLEFT همراه با PDA وASD و همانژیومای زیر زبانی درنوزادان

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دکتر هادی سماعی

Sternal Cleft, PDA, ASD and Subglottic Hemangioma in neonate

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Abstract: We describe the rare case of a3day –old- girl with a sternal cleft (Fig1), PDA, ASD and subglottic hemangioma. She underwent a surgical repair of sternal cleft and treatment of hemangioma with Perdnisolon. But she was readmitted due to respiratory distress and apnea in 8 months old. She underwent a tracheostomy and after one week she underwent a laryngoscopy and bronchoscopy. That identified a subglottic hemangiomawas treated with theKTP532 laser. Now she is 3years and 10 months old with, no respiratory distress or any recurrent of hemangioma.

Introduction: Sternal cleft is a rare visually dramatic congenital anomaly .It result from failure of fusion of the 2 lateral mesodermal sterna bars by the 8 weeks of gestation. Subglottic hemangiomas are rare vascular malformations that enlarge from shortly after birth until 8-18 months of age, when spontaneous involution begins. Complete regression usually occurs 5-8 years of age2. Numerous treatment modalities have been advocated most commonly tracheostomy, systemic steroids, carbondioxide laser, or interferon alfa-2a.Any physically distractive treatment may lead to permanent subglotic stenosis.

Case Report: A full–term neonate was post nataly diagnosis with a u- shaped cleft of the upper half of the sternum, with central area of ulcerated skin overlying the visible pulsating heart. She was born on the 40^{th} week gestational age to 31 year olds gravid 2, para2 mother and a 38 year old father an uneventful pregnancy by caesarean section. Birth weight was 2670grand length was 48 cm. There was no history of maternal infection or medication during early pregnancy .The family history was unremarkable and parent was not consanguineous.

Dimensional and Doppler echocardiography showed a sternal cleft, normal heart anatomy, active PDA, ASD and normal aortic arch.Ultrasonography of abdomen was normal.

In laboratory investigation revealed kelebsiela pneumonia and entro bacter spp in tracheal specimen. Due to culture and sensitivity of germs the patient treated by Vancomycin, Amikacin and Cefotaxime. The patient underwent surgery at 3 day of age in order to repair of sternal cleft. The necrotic fibrous tissue in the sternal area was completely resected without open the pericardium. Fasciocutaneous flaps were raised on either side, and approximated two sternal bar with 2/0 PDS and covered it with fasciocutaneous flaps. Sterna cleft closed and cardiac pulsation were no more visible through the defect after the reconstruction. At the age of 8 months she was readmitted because of the severe respiratory distress, stridor and sternal notch retraction, apnea and hemangioma in the lower lip had also extended to the oral mucosa and right parotid region. Spiral CT-scan from skull and larynx revealed partial and subglottic hemangiomas. She underwent a tracheotomy and after one week; she underwent a laryngoscopy and bronchoscopy and subglottic hemangioma treated with KTP532 laser. The functional and cosmetic result was good after complete healing.

گزارش یک مورد اکتوپی کوردیس

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True Ectopia Cordis; Report of a Rare Case

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True ectopia cordis is an extremely rare congenital anomaly in which the heart has no overlying somatic structures. There are also chest and abdominal wall defect. The incidence is 5.5-7.9 per 1 million live births. The prognosis is poor and depends on the severity of the intracardiac malformations. Surgical repair is a challenge with only few survivors. Although prenatal diagnosis reported as early as the first trimester, our patient was born full term on elective caesarean section. We report a successful surgery of ectopia cordis that the heart was naked, siting on the chest and the apex pointed toward the chin. The sternum was entirely deficient except a rim of manubrium. Also there was upper abdominal wall defect. The heart was successfully covered by two skin flap. A small piece of Gore-tex patch sutured to the edges of defect. Postoperative echocardiography revealed tetralogy of Fallot.

یک روش جدید در کلستاز کبدی فامیلی برای درمان خارش سخت

Partial Internal Biliary Diversion For The Treatment of Intractable Pruritus in Children With Progressive Familial Intrahepatic Cholestasis: Introduction of a New technique

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Introduction: Chronic intrahepatic cholestasis (Byler's disease) is associated with severe pruritus that is often refractory to maximal medical management and leads to significantly impaired quality of life. No known medical therapy has been successful in lowering levels of bile salts. Numerous procedures have been proposed for biliary diversion, but there is not any standard approach.

Aim: The aim of the study was to describe the initial experience with a novel approach to the surgical treatment of progressive familial intrahepatic cholestasis (PFIC).

Methods: The patient was 10 year old girl underwent partial internal biliary diversion to treat incontrollable pruritus associated with PFIC. The surgical technique involved the creation of an isolated appendicular conduit, anastomosed distally in a termino-lateral fashion to the gallbladder (cholecystoappendicostomy). This operation combines the advantages of partially diverting the biliary flow from the enterohepatic cycle, avoiding an external biliary fistula and use of just one anastomosis.

Results: The patient had complete resolution of her pruritus. No complications were observed related to this operation. There has been no clinical evidence of progression of liver disease.

Conclusions: cholecystoappendicostomy as a technique for Partial internal biliary diversion is an effective surgical option for the treatment of PFIC. Long-term follow-up is necessary to evaluate late results and eventual complications of this approach

Bacille Calmette-Guérin lymphadenitis

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Bacille Calmette-Guérin (BCG) lymphadenitis is the most common complication of BCG vaccination. Two forms of BCG lymphadenitis can be recognized in its natural course—simple or non-suppurative lymphadenitis, which usually regresses spontaneously over a period of few weeks, and suppurative BCG lymphadenitis distinguished by the development of fluctuations in the swelling, with erythema and oedema of overlying skin. Healing in suppurative glands occurs through spontaneous perforation and sinus formation, followed by closure of the sinus by cicatrisation. Non-suppurative BCG lymphadenitis is best managed with expectant follow ups only, because medical treatment with erythromycin or antituberculous drugs do not hasten the regression or prevent development of suppuration. Suppurative BCG lymphadenitis may be treated by needle aspiration to hasten resolution and prevent spontaneous perforation and sinus formation. Surgical excision is rarely needed and is meant for cases of failed needle aspiration or for draining BCG nodes.

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Congenital lobar emphysema in a 5 years experience

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Introductions and Aim: congenital lobar emphysema (CLE) is a rare congenital anomaly by over inflation of a pulmonary lobe. We review the clinical result of our patients with CLE.Highlighting the results of operation and complications.

Patients and methods: In a retrospective study from 2007-2012 we evaluated 22 patients with CLE. Variable collected included: age, sex, clinical manifestation and radiological diagnostic methods. Pre and post operative complications and type of treatment

Result: Twenty two patient, male to female ratio (16 male 73%, 8female 27%). The mean age of males and females patients were (8 ± 4.7 and 4±7.2 months) respectively. The main diagnostic methods were CXR and chest CT, but a few of them have only chest x ray. The most affected lobe was left upper lobe (54%), associated anomaly were seen in two patients. All of the patients under went thoracotomy, Mortality rate 0% one patient was bilobar involvement.

Conclusion: This study confirms that the careful evaluation and treatment in equipped hospital and pediatric anesthesiologist directly affect on the result and survival.

سکستراسیون ریوی (گزارش ۵ مورد)

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Pulmonary Sequestration (reporting 5 cases)

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Background: Pulmonary sequestration is a cystic or solid mass composed with none functionally primitive tissue. It does not communicate with the tracheobronchial tree and has anomalous systemic

Blood supply. It is a kind of thoracic malformation and may present as a lung infection in physical examination and chest imaging. Its blood supply is from systemic circulation rather than pulmonary circulation. Multiple feeding vessels may be present in 15-20 % cases. The two forms of pulmonary sequestrations are; intrapulmonary, which is surrounded by normal lung tissue, and extrapulmonary, which has its own plural investment. Other congenital malformations may present in this anomaly.

Cases presentation: In this report we discuss about five patients aged between 4 days to 3 years, referred for CDH repair, recurrent and chronic pneumonia. There were 2 boys and 3 girls; among them one patient was referring for CDH repair and four patients for pneumonia. Diagnosis was based upon history, physical examination, imaging and intra operative findings.

Results: During CDH repair we found one patient with extrapulmonary sequestration that resection of sequestration and repair of CDH was done, and four patients with intrapulmonary sequestration. All sequestrations have been in the left lower lobe and all were completely resected with lobectomy. Sequestration supplied with aberrant vessels that originated from aorta which were ligated and divided. There was no mortality in our cases. Pulmonary sequestration is a relatively rare congenital anomaly, mostly presented with recurrent or chronic pneumonia. This anomaly is cured after surgery.

Conclusion: In surgery we must focus on aberrant blood supply that is originated from systemic circulation. In chronic or recurrent pneumonia, sequestration must be suggested in differential diagnosis.

تظاهرات حاد وتاخیری ضایعات مادرزادی کیستی ریه، گزارش یک مورد و بررسی متون

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Late and Acute presentation of congenital cystic lesions of lung: Report of Case and Review of Literatures.

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Introductions and Aim: Congenital cystic lesions of lungs are uncommon, but have similar embryologic and clinical characteristics. Some of these lesions have late and acute presentation with unusual clinical manifestation.

Case report: 7 months old girl that presented to pediatric emergency department with respiratory distress and mild cyanosis. On physical examination child was tachypenic and restless with decreased breathing sound in left thorax. In chest X-ray there was huge pneumothorax in left thorax with shifting of mediastinum. Patient was admitted immediately to PICU for respiratory support. Surgical consult was done. Resident surgeon asked the pediatric surgeon for chest tube insertion. Evaluation of patient by pediatric surgeon showed her respiratory distress is mild and her O2 saturation was 93% and her vital signs are stable. Chest tube insertion cancelled and patient kept in PICU under close observation for more investigation. Chest CT was done in the next day that confirmed cystic lesion of left lung. Left thoracotomy was done that showed a single huge lobulated cystic lesion from lingula lobe of left lung that inflate with inspiration and compressed the normal lung. Resection of cyst was done. The left lung expanded well. Patient has had uneventful recovery and discharged from hospital in a few days.

Conclusion: In patient under 1 year old, cystic lesions were discovered by respiratory distress; and in patients over 1 year old signs of infection were the most important clinical features. Early recognition of these relatively rare congenital cystic lung lesions would lead to the immediate, proper surgical intervention.

آسپیراسیون غیر معمول جسم خارجی

دکترسهیل بن رضوی، دکتر محمد بمانیان

Unusual foreign body aspiration

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A 1.5 Y/O girl a case of brooch (pin) aspiration since last night, consulted after two unsuccessful try for removal of that by ENT specialist.

General condition of baby was not good and she was in respiratory distress. Change in location and shape of pin was noticed on X-rays. Baby pushed to OR and successful removal of pin was done. General condition of baby was good.

کیست بورونکوژنیک در یک بیمار با آسم

Bronchogenic Cyst in a patient with Asthma

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Difficult to treat asthma is an asthma syndrome that brings in our mind other differentials. Mediastinal masses are not common findings, but are important variables. Bronchogenic cyst is a congenital anomaly of the foregut that is typically found in the mediastinum and diagnosed accidentally. We present a 4-year-old girl with allergic asthma that began at 8-months of age and finally a bronchogenic cyst was detected in this patient. The patient had history of asthma since she was eight months old. She had a history of several asthma attacks which had partly responded to asthma management.

During the last episodes of asthma attacks, she was hospitalized in Pediatric Intensive Care Unit. Imaging studies showed a 4×3 cm mass in the posterior part of the thoracic cavity that had led to tracheal narrowing was found for which the patient underwent thoracotomy and in surgical exploration a cyst that had compressed the thoracic trachea. Pathological examination of the cyst revealed a bronchogenic cyst. Bronchogenic cyst is an uncommon developmental abnormality but in a patient with obstructive pattern of airways it should be considered in differential diagnosis of asthma, especially if the asthma management is not successful.

دکتر مسعود ناظم، دکتر عبدالحمید عموئی، دکتر محمد عیدی

Duplication of Cervical oesophagus: a case report and review of Literatures

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Introductions and Aim: Foregut duplication is commonly found in the posterior mediastinum. 10-20% of these anomalies are associated with oesophageal duplication. It can occur in all parts of oesophageal length. Although duplication of cervical oesophagus has been previously reported, but a majority of them were found in thoracic oesophagus, infants with oesophageal duplication usually manifested by respiratory distress or asymptomatic thoracic mass, casually, detected in X-ray. A 7-month-old infant weighing about 7.5 kg, with the signboard of respiratory distress, fever and nutritional intolerance was admitted to our hospital. Physical examination showed dehydration, stridor, tachypnoea, intercostal retraction and neck stiffness. Plain chest radiogram showed dubious cystic mass in the distal neck. Cervical and chest computed tomography scan showed neck cystic lesion disseminated to posterior mediastinum, probably propagated to the respiratory system. Cystic lesion connected to oesophagus was partially resected, oesophagus was repaired and remaining mucous of a cyst was removed, then gastrostomy tube was applied.

Although cervical oesophageal duplication cysts are rare, but they must be considered as one of the differential diagnoses of cervical mass with respiratory distress in infants

درمان کیست تیروگلوس عود شده با فیبرین گلو و فاکتور رشد

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Treatment of Recurrence of Thyroglosalsist with: Fibrin Glue and growth Factor

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Introductions and Aim: of the Study: Congenital Thyro-glosal cyst needs to surgical correction. Recurrence is unusual but its treatment is difficult and accompaniments with greater scar, cervical structures damage and else. We have used an alternative treatment which using riched fibrin glue with growth factor (RFGF).

Methods: Three cases who previous surgery for thyro-glosal cyst reconstruction had presented with saliva secretions from incision site. They have operated 14-20 and 60 days before recurrence.

We did cervical ultrasonography for cases. Cysts size was 1×4cm, 1.5×1.5cm and 1.5×2 cm. We aspirated cyst contents and injected RFGF as 0.5ml more than aspirated volume into cyst. Saliva secretions stopped immediately after injection. We followed them 1, 3, 6, 12 and 24 month later.

Main results: No findings of cystic lesion were on ultrasonographic exam which have done on 3 to 24 month after surgery.

Conclusion: Recurrence is an unusual but disaster complication of thyro-glosal cyst surgery. It may be accompaniment with some events such as salivary fistula, superimposing with infection, neck mass .Reoperation could be difficult due to tissue adhesion tenacity. Then that is reasonable to use some techniques that no needs to surgery. Fibrin glue packs cyst and growth factor motivates cell proliferation. Since use of riched fibrin glue with growth factor is accompaniment with cosmetic acceptance, no leakage of cyst secretion and no need to reoperation we advise replace corrective surgery with this technique.

روش داخل جداری لیلی بهترین روش برای اکسزیون کیست کولدوک

Intramural (Lilly’s) method is the best procedure for resection of choledochal cysts

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Introduction: Cystic dilatation of common bile duct (CBD) or choledochal cyst has two clinical presentations, infantile and adult form. About 90- 95 percent of choledochal cysts are type I that is fusiform dilatation of extra hepatic billiary dilatation only. Choledochal cyst has a thick wall with dense connective tissue and because of chronic inflammation adhesion to surrounding tissue is severe. Despite of multiple procedure for operation because of high rate of complications and especially carcinoma, today, complete excision of the cyst and billiary bypass have become the treatment of choice. We present three adult type cases of choledochal cyst that presented with different symptoms and operated with intramural (Lilly’s) method.

Aims: Because of difficulty in complete resection of cyst and complication we suggest lily's method is the best and primary method for operation.

Methods and Result: Two female 9 Y/O & 1 Y/O and one male 2.5 Y/O presented with abdominal pain intermittent jaundice and with diagnosis of choledochal cyst candidate for operation: in all patients during operations firstly we tried to resects en-bloc cyst with, gall bladder extrahepatic billray ducts, but because of severe adhesion, bleeding and possible damage to surrounding important organs we changed operation to intramural (Lilly’s) method that was very easier with no complications.

Conclusion: Intramural (Lilly’s) operation for resection of choledochal cyst is the best method because it is time saving- resection of pathology is simple, lower rate of bleeding and complication at operation and good outcome.

Mirizzi's syndrome در کودکان با تابلوی کیست کولدوک: گزارش دو مورد

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Mirizzi’s syndrome mimicking choledochal cyst: Report of two cases in children

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Introduction: Mirizzi’s syndrome is a rare cause of acquired hyperbilirubinemia that tends to affect older people more often. It is a type of obstructive jaundice that results from choledochal compression either by the stone or from fibrosis caused by cholecystitis. Obstructive jaundice and subsequent hyperbilirubinemia deserve a wide range of differential diagnosis such as choledochal cysts which could potentially delay the diagnosis of Mirizzi’s syndrome.

Case report: We report two cases of Mirizzi’s syndrome in children. The first child was an 18 months old girl and the second was a four years old one. Both of the cases were diagnosed to have a choledochal cyst based on preoperative radiologic examinations. The patients’ clinical presentations and laboratory findings were also suggestive of the diagnosis of a choledochal cyst. Both of the patients had been candidates of intraoperative cholangiography. The cholangiographic examination revealed a dilated common bile duct (CBD) together with a dilated common hepatic duct (CHD) in both of the patients. After an appropriate surgical exposure, we found that both patients have had a dilated cholecyst with compression effect on CBD and CHD with subsequent obstructive jaundice and direct hyperbilirubinemia suggestive of Mirizzi’s syndrome. The primary clinical and radiological presentations of both patients were also justifiable with the intraoperative findings. The surgical explorations ruled out any cystic lesions in CBDs of the presented cases.

Conclusion: Mirizzi’s syndrome is a rare cause of acquired jaundice in pediatric surgery and its diagnosis is delayed many times. It should be considered in the differential diagnosis of obstructive hyperbilirubinemia in infants and children. Obstructive jaundice caused by Mirizzi’s syndrome may mimic clinical and radiologic pattern of a choledochal cyst. Therefore, this syndrome should be in mind in a child supposed to have a choledochal cyst.

پی آمد تغذیه زودرس در کودکان با آناستوموز روده یک مطالعه مقدماتی

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Outcome of early oral feeding in children undergoing intestinal anastomosis: a preliminary study

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Introduction: Intestinal anastomosis is followed by NPO times in adult and pediatric population. It is assumed that NPO times maintain the integrity of anastomosis and prevents early and late complications. We conducted a study to assess whether early feeding could complicate the anastomosis or not.

Method and materials: We studied 40 consecutive children undergoing intestinal anastomosis. We randomly divided the study patients into two groups of early and routine postoperative oral feeding. We initiated oral feeding on the third postoperative day for the patients in early feeding group; while we initiated it on the fifth day for the patients in routine group. Mean days of admission, initiation of intestinal peristaltism, gas passage, postoperative ileus, and postoperative complications were compared between two groups. Postoperative complications included leak of anastomosis, hematoma, abscess formation, and peritonitis.

Results: The mean age of study patients was 6.3 ± 7.5 years of age. Thirty patients (75%) were male and ten patients (25%) were female. Background diseases necessitating intestinal anastomosis were Hirschsprung's disease, imperforate anus, gangrenous intussusceptions, ileal atresia, and intestinal obstruction. Mean admission time in early feeding and routine feeding groups were 4.4 ± 0.7 and 5.5 ± 0.7 days respectively ($p < 0.001$). The mean day of initiation of intestinal peristaltism was 2.9 ± 0.7 in early feeding group; while it was 3.1 ± 0.7 in routine feeding group ($p > 0.001$). There was not also any significant difference in postoperative ileus between two groups ($p > 0.001$). Postoperative complications did not occur in any of the study patients.

Conclusion: Early initiation of oral feeding in children undergoing intestinal anastomosis deserves the integrity of anastomosis similar to routine NPO days. Early feeding does not complicate gastrointestinal anastomosis and it should be considered in pediatric bariatric surgery.

یک مورد نادر SIRENENOMELIA, MERMAID و بررسی متون

Sirenenomelia, the Mermaid Syndrome a Rare Case Report and Review of Literacture

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Sirenomelia, the Mermaid syndrome is an extremely rare and lethal congenital anomaly with incidence of to 1 in 60-100000 births. It is characterized by complete fusion of the lower limbs associated with external genitalia absence, renal agenesis and anorectal defects. Sirenomelia represents the most severe of caudal regression syndrome. Although prenatal diagnosis reported as early as the first trimester, we present an infant delivered from a 19 – year- old primigravida at 33 weeks gestation. A single, dysmorphic lower limb, agenesis of external urogenital and anorectol atresia were detected at delivery. X- Ray

(Baby gram) and autopsy revealed other anomalies in the latter.

انحراف داخلی صفراوی در کودکان با PFIHC

دکتر حمید فروتن، دکتر علی بهادر، دکتر محسن سلطان نجفی

Internal Biliary diversion in children with PFIHC

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Introduction and Purpose: PFIC remain a difficult problem for both physician and parent of patients. The aim of the present study was survey of effect a new surgical approach (internal drainage) for progressive familial intrahepatic cholestasis disease emphasizing the indications for surgery, surgical techniques complication and results.

Method: In our study ten patient with PFIC were operated in our center. These cases were reviewed for treatment response and determinate level of bilirubin (total, direct), score of pruritus described by Whitington and Whitington and primary or early complication which was happened in during time of fallow up.

Result: Ten patients presented with severe pruritus underwent surgical treatment (9 patients had PFIC, one patient known case of Alagille Syndrome), all patient had elevated bilirubin before surgical intervention and the diagnosis was documented by liver biopsy and (role out) another disorder. There were 8 patients clinically and paracilinically good response to surgery. There were significant improvements in pruritus score and level of serum bilirubin. Two patients were not responded by this surgical technique.

Conclusion: We recommended internal drainage in the treatment of PFIC who was suffering from sever pruritus (itching).

گزارش یک مورد تومور نوروآندوکرین پانکراس در یک کودک سه ساله

دکتر سید جواد نصیری، دکتر شیرین سیاح فر، دکتر میترا مهرآزما، دکتر مهناز صادقیان

Report of a case of Neuroendocrine Pancereatic Tumor in a three years old child

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Neuroendocrine tumors of the pancreas are rare specifically in pediatric age group. One of the Symptoms of these tumors is jaundice which may be misleading especially in countries with high prevalence of hepatitis A .We report a 3.5- year -old girl with four weeks of icterus with primary misdiagnosis of hepatitis A , differential diagnosis of fusiform choledochal cyst and final diagnosis of well differentiated low grade neuroendocrine carcinoma of the pancreas. With our best knowledge this is the first report of this tumor in under -5 -year-group.

مطاله کودکان با نزیدیوبلاستوزیس در اهواز

دکتر شهنام عسکر پور، دکتر مهران پیوسته

Nesidioblastosis in children: A study from Ahvaz

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Introduction: Persistent hyperinsulinemic hypoglycemia of infancy (PHHI) is the most common cause of persistent hypoglycemia in infants. Consists of a group of distinct genetic disorders causing dysregulation of insulin secretion, the current standard treatment is near total pancreatectomy. However, the long-term outcome following surgery needs further attention. Recurrent episodes of hyperinsulinemic hypoglycemia may expose to high risk of brain damage. Hypoglycemias are diagnosed because of seizures or any other neurological symptoms, in the neonatal period or later, usually within the first two years of life. Near-total pancreatectomy is the procedure of choice for diffuse form. Oral Diazoxide is a first line treatment. In case of unresponsiveness to this treatment, somatostatin analogues and calcium antagonists may be added**.**

Material and Methods: A retrospective review of 9 patients diagnosed with CHI between 2000 and 2011 was performed .all patients underwent near total pancreatectomy. Data were collected on age, gender, clinical presentation, medical and surgical management, and complications**.**

Result: 6 patients developed symptoms within 72 hours after birth (early-onset).Convulsion, cyanosis, and lethargy; refusing milk sucking, irritability and sweating were common symptoms. The laboratory findings displayed persistent hypoglycemia and hyperinsulinism in all of the 9 infants. The glucagon test showed positive and no urine ketones were detected in all of the 9 infants. 6 presented with seizures. All were initially treated with Diazoxide, and all patients required near total pancreatectomy .4 developed permanent diabetes mellitus

Conclusion: The aim of management of PHHI should be preventing mental retardation due to severe and persistent hypoglycaemia, and on the other hand to avoid the induction of diabetes mellitus following surgery. Satisfactory outcome depends on early effective medical treatment and judicious use of surgery by an experienced operator 95% pancreatectomy is an effective treatment modality in PHHI not responding to medical treatment. In our experience; recurrence of hypoglycemia following surgery responds well to medical treatment. There is a high risk of secondary DM in these children; however, the serious risk of brain damage and neurological abnormalities could be avoided by early surgical intervention

اخلاق در جراحی

دکتر ایرج فاضل

Surgical Ethics

Fazel I

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Duties of surgeons to patients: A surgeon must: 1- Maintain the highest professional standard and practice his profession without a profit motive. 2- Be obligated to preserve human life. 3- Be loyal to his patients. 4- Call for a second opinion if a certain type of treatment is out of his ability.5- Maintain confidentiality on his knowledge of patients. 6- Give emergency care where needed as a duty unless others are able and willing to administer such care. **Four areas of communication: 1- What to tell:** Tell the truth if at all possible, use clear non-puzzling language, tell the truth calmly, sit at the same level as the person to whom you are speaking, discuss treatment options.**2-When to tell:** When all relevant results are available try to tell the patient and relatives as soon as possible.**3-Whom to tell:** Tell the patient, use carefulness when the prognosis is very poor, permit the patient to ask questions, he has a right to know what is happening to him, discuss the clinical aspects of the diagnosis with the closest relatives, reassure them that a truthful approach will permit maximum co-operation from the patient and also justify future admissions, treatments, or continued follow-up at hospital.**4-Where to tell**: Speak to the patient or his relatives in privacy not in the corridor. If in the open ward, draw the screens and ask the nurse responsible for the patient's care to accompany you. **Informed consent:** For agreement to account as consent to treatment, patients need to be given appropriate and accurate information about: Their condition and the reasons why it warrants surgery, what type of surgery is proposed and how it might correct their condition, what the proposed surgery requires in practice. the anticipated prognosis of the proposed surgery, the expected side-effects of the proposed surgery, the unexpected hazards of the proposed surgery, any alternative and potentially successful treatments for their condition other than the proposed surgery, along with similar information about these, the consequences of no treatment at all.

Who should be responsible for taking the consent? Ideally, the person providing the treatment or a delegated person who is sufficiently trained and qualified and has sufficient knowledge of the proposed procedure. **Who should give the consent?** The patient, the presence of relatives and friends can sometimes be useful. However, no one else may make decision on behalf of a competent adult. If there are language difficulties, the use of an independent interpreter should ensure that the correct information is delivered to the patient. If a patient asks you withhold information or make decision on their behalf, you should stress the importance of them knowing the different options, and you should still provide the basic information about the treatment. **Duties of surgeons to one another:** A surgeon must:1- Behave respectfully and professionally towards colleagues. 2- Not attempt to or succeed in attracting patients from surgical colleagues. **Unethical Practice:** 1- Self advertisement.2- Collaboration in medical practice where clinical independence is not maintained.3- Receiving monies, other than proper professional fees.4- Acts or advice which could. weaken the mental or physical status of a person and which could result in profit of some kind for the surgeon.**Warning:**1- Beware of new discoveries and techniques unless they are properly tried and tested.2- Give certification or testify only to that which you can confirm personally.

کیستهای معده دوپلیکاسیون فورگات

دکتر عبدالحمید عمویی، دکتر محمد وفایی

Foregut Duplication cysts of the stomach

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Introduction: The term "gastric duplication" implies the presence of gastrointestinal mucosa (usually gastric, but may be small intestinal or colonic), whereas the term "foregut duplication" is preferred when pseudo stratified ciliated epithelium predominates. Duplication cyst of the stomach with pseudo stratified columnar ciliated epithelium is extremely rare

Case Report: Our patient9-year male presented with massive upper G.I bleeding. Abdominal examination and routine laboratory tests except low Hb and Hct were normal. The patient was resuscitated with serum ringer and packed red blood cells. The patient underwent upper G.I endoscopy, which revealed an extrinsic compression at fundus of stomach with big ulcer (3 cm diameter) in central portion, with active bleeding and normal-appearing gastric and duodenal mucosa. Gross examination of the surgical specimen revealed a unilocular cyst measuring 10cm in diameter with a grey-white, rubbery wall. Histopathological examination showed a cyst lined by pseudo-stratified, ciliated, columnar epithelium with a partial lining of smooth muscle bundles

Discussion: Alimentary tract duplications are rare congenital anomalies that can occur anywhere along the alimentary tract from the tongue to the anus with the most common site, the ileum. Gastric duplication cysts as a subset make up approximately 4% of all alimentary tract duplications with a review of 80 cases demonstrating a 2:1 female-to-male ratio. Duplication cyst of the stomach with pseudo stratified columnar ciliated epithelium is extremely rare clinically, gastric duplication cysts can present with vague abdominal pain, vomiting and occasionally a palpable abdominal mass but are usually asymptomatic and, therefore, difficult to diagnose. Due to surrounding gastric inflammation and ulceration, marked haematemesis, melaena stool are the only presenting symptoms, as was the case in our patient. A high index of suspicion is needed when dealing with the evaluation of haematemesis of unknown cause in children and experience as well as review of endoscopy may be required even when using the gold standard.

گزارش دومورد PDPV : Preduodenal portal vein

دکتر حلیم بردی طعنه، دکتر غزاله دزیانی

PDPV :Two case of preduodenal portal vein

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PDPV is rare developmental anomaly. Injury to these structures because failure to recognize them during operations way.

Result in thrombosis or hemorrhage. We recently encountered this anomaly. Twice: once in a neonate with failure to pass meconium and history of maternal Polyhydramnios and another one in a 7 month old infant with recurrent biliary vomiting and upper abdominal distention.

Case 1: A 10 day –old female neonate weighting 2/4 kg born to G4P4L3 Mother at 35 weeks of gestation by cesarean section elsewhere. She was born with tachypenia and admitted in the hospital and was treated with antibiotics, after ten days she was referred for complaints of failure to pass meconium and upper abdominal distention. Polyhydramnios was reported on the antenatal ultrasonography in the third trimester of pregnancy and she got GDM and Preeclampsia.

At second admission, the patient was quiet and vital sign were normal. There was minimal upper abdominal distention; she was icteric from face to abdomen. Gastric peristalsis was visible. The patient was managed with intravenous fluids and antibiotics. Naso-gastric aspirates were bilious. Blood biochemistry was normal except bilirubin. Total bilirubin was high. Whole laboratory test for sepsis were reported normal.

Upper GI study (fluoroscopy) was done and oral contrast media entered through NG tube but it did not pass through duodenum and abdominal distention was seen. After that laparotomy was undertaken that revealed a prominent PDPV obstruction between the first and second part of duodenum **(d1 and d2**).

Duodenojejunostomy was done. After operation, she received antibiotics and NG tube was fixed for her. After 7 days, NG tube was disconnected and started breast feeding. After 14 days, she discharged with well general condition.

Case 2: A 7 month-old female weighting 6 kg born to G1P1L1.she was referred for complaints of biliary vomiting after take the semisolid diet upper abdominal distention. The patient was managed with intravenous fluid and antibiotic. Naso-gastric aspirates were bilious. Upper GI study was done and was reported partial obstruction in the second part of duodenum. Laparotomy was undertaken that revealed a prominent PDPV obstructing between the first and second part of duodenum. After 7 days she discharged with well general condition

استنوز مادرزادی کولون یک گزارش مورد

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Congenital colonic stenosis a case report

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Introductions: Colonic atresia and stenosis are the rare entities with an incidence (colonic atresia) of 1 in 20,000 live births and comprise about 1.8-15% of intestinal atresias. The colonic stenosis is even rarer and to date not more than 10 cases have been reported in the English literature. (1) We describe the case of a 2-month-old female infant coming to our attention because of an intestinal sub occlusion due to a congenital colonic stenosis of the colon.

Case report: A 2-month-old female infant presented with abdominal distension and vomiting. She had these symptoms from twenty-days of birth and one week before this admission, she was admitted because of premarurity (30_31 weeks), icterus and sepsis. On clinical examination Abdomen was distended with visible bowel loops. Bowel sounds were absent. Digital rectal examination revealed empty rectum. Nasogastric tube was passed and intravenous fluids and antibiotics started and surgery planned.

Abdominal radiograph showed distended abdominal loop. During contrast enema sigmoid colon filled with contrast and a narrowing was seen in distal of descending and sigmoid colon; Suspecting an organic intestinal obstruction.(Figure 2)An explorative laparotomy was deemed necessary and between distal of descending and sigmoid colon a stenosis was found. (Figure 3) This portion of colon was resected and an end to end primary colo-colic anastomosis performed. The post operative recovery was uneventful. Naso-gastric tube was removed on 4th post operative day and patient started orally and discharged. According to literature review and our experiment while congenital colonic stenosis is rare but it seems that can cause complete or partial intestinal obstruction, not only in newborns but also throughout the first year of life

نتایج ترمیم گاستروشیزیز در مرکز طبی کودکان

دکتر جواد احمدی، دکتر بهار اشجعی

A study of Gasrochisis repair in Children's Hospital Center

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Introduction: During past decades the mortality rate of gastrostschisis was reduced from 90% to almost 13% .These advantages are due to better mechanical ventilation and parenteral nutrition and staged repair by using prosthetic materials.{, #4}.Other factors that can change the mortality rate of gastroschisis are prematurity and other congenital malformations and disparity between abdominal cavity and eviscerated organs.{, #1}.in general the mortality rate of gastroschisis is reduced during forty years ago.{, #3}

Materials and Methods: This is a retrospective study that is performed in children's medical center of Tehran University of Medical Sciences. All neonates who were admitted in NICU due to gastroschisis in past 10 years and needed to silo for staged repair were included in our study. We reported the rate of coexisted congenital anomalies and the complication rates of surgical and medical managements. Staged operations were done in 42 patients based on discrepancy between peritoneal cavity and eviscerated organs. We used a sterile urine bag as silo in these patients and every 24-48 hours we made the bag smaller by wrapping the bag and suturing the top of it.

Result: We had 42 infants who were admitted in NICU in ten years period and undergone staged repair of gastroschisis. Coexisted congenital anomalies except malrotation and incomplete small bowel fixation were detected in 19 patients. These anomalies were biliary atresia in 2 patients, jejunal atresia in 2 patients, ileal atresia in 1 patient, bladder extrophy in 1 patient, congenital heart disease in 3 patients, undessendent testis in 10 patients, urinary anomalies in 3 patients, inguinal hernia in 12 patients and urinary tract anomalies in 2 patients. Total mortality rate was 5 in 42 cases. Cardiac anomalies in 2 patients, sepsis in 2 patients and pneumonia in one patient were the cardinal cause of death.

Conclusion: Mortality rate due to gastroschisis is reported from 6% to 20-35% in different centers and it is highly dependent to other congenital anomalies, the time of evisceration of bowel, the technique of facial closure and medical care.

واسکولیت روده باریک

Vasculitis of small bowel

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A 5 Y/O boy with sever colicky abdominal pain since 3 days ago reffered by pediatric infectious specialist for surgical intervention. Results of US and CT scan were NL, lab data was no specific. laparatomy was done with finding of multiple area of segmental thickening and Hemorrhagic pattern in a length of 5-7 cm . One of segments was resected and anastomosis was done other finding was NL.

3days after operation some purpura was appeared in body of patient. Result of pathology was in favor of H.S.P.

خونریزی شدید گوارشی با علت دوپلیکاسیون روده

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Sever GI bleeding due to intestinal duplication

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A 10 months old infant had been admitted at Abouzar Children's Hospital with severe lower GI bleeding. In the first laboratory examination patient's hemoglobin were 8.5 which gradually dropped to 5.3. After receiving crystalloid fluid and blood transfusion upper GI endoscopy was performed which was normal. In HIDA scan duplication of intestine with ectopic gastric mucosa was reported. The patient underwent laparatomy surgery. Duplication of intestine was present at ileum, 70-80cm away from ileosecal valve. Resection and end to end anastomosis was performed and patient discharged uneventfully.

دوپلیکاسیون دستگاه گوارش

دکتر فرید اسکندری، دکتر هوشنگ پورنگ، دکتر منصور ملائیان، دکتر هدایت الله نحوی،
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Alimentary Tract Duplication

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Alimentary tract duplication is a rare congenital disease which is presented as a mass adjacent to different parts of the alimentary tract from mouth to anus. It is either presented with GI bleeding or obstruction, abdominal distention, abdominal mass, or sometimes symptomless which discovered by accident. The majority of duplications are diagnosed by the age of two years, but may also be discovered before birth by ultrasonography.

There are multiple theories to explain GI duplication occurrence, like as "partial or abortive twining", "split notochord", and" diverticula and canalization defects" but none of them can account for all the known variants.

Duplications are benign malformations; however due to presence of ectopic gastric mucosa there might be a chance of peptic ulcer and malignancy in rare cases. They often located on the mesenteric side and share the same vascular structures of native alimentary tract. These lesions may be either cystic or tubular, and ileum is the most common enteric site of duplication

Radical resection is considered to be the best treatment but it is not always possible to do solely without resection of adjacent bowel. Treatment of large thoracoabdominal or long abdominal duplications can be challenging. The main goal of surgical treatment is to clear the symptoms and prevent any form of recurrence. Prognosis is good, yet depending on severity of the disease, pathogenicity, morbidity and mortality of the associated anomalies.

This paper introduces some patients whom were finally diagnosed with different parts of the alimentary tract duplications including pylorus, duodenum, small intestine, and colon, and being treated in Bahrami Children's Hospital

تظاهردوپلیکاسیون کامل کولون با توده لگنی و پرولاپس رکتوم

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Complete colonic duplication presented as pelvic mass and rectocele

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Purpose: complete colonic duplication is very rare anomalies that may have different presentation according its location and size. Complete colorectal duplication can occur in 15% of gastrointestinal duplication. This report presents a complete colorectal duplication that presented as chronic constipation, pelvic mass and rectocele.

Case Report: 2 year old boy presented to clinic with abdominal protrusion, difficulty to defecate, chronic constipation and prolapse of mucosa covered bulging (rectocele) since age 6 months. On examination abdomen was soft with palpable pelvic mass with doughy consistency. Rectal exam confirmed prerectal mass with soft consistency. Plain abdominal X-ray showed dilated bowel loops and soft tissue pelvic mass that pushed rectum anteriorly and cervical hemi-vertebra. Barium enema and abdominal and pelvic CT confirmed the same finding. US revealed absence of right kidney that confirmed by DMSA scan, VCUG was normal. Laparatomy was performed, there was a complete colorectal duplication with one blinded end and accumulation of huge amount of stool that protruded as mucosa covered bulging during defecation (rectocele). The duplicated colon has common mesentery and resection of one part was impossible. The mucosal web was resected and the two ends of duplicated colon were fenestrated to each other and opened to common anal canal and all fecal impaction was evacuated and patient had very good post operation recovery and discharged from hospital in a week time. In two year follow up has normal defecation and good weight gain without any problem with duplicated colon.

Conclusion: The Side to side total colorectal duplication may associated with urinary and vertebral anomalies and can be treated with simple resection of distal common wall, fenestration.

یک متد تازه وموثر در جلوگیری ازایجاد و لولوس روده بعداز عمل در Apple Peel Jejunal Atresia

دکتر احمد محمدی پور

A new and useful method for prevention of postoperative volvulus in Apple Peel Jejunal Atresia (Christmas – tree deformity)

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Type III(b) Atresia (apple peel, Chrismas tree or Maypole deformity) consist of a proximal jejunal atresia, absence of the superior mesenteric artery beyond the origin of middle colic branch, agenesis of the dorsal mesentery, significant loss of intestinal length and large mesenteric defect .The decompressed distal small bowel lies free in abdomen and assumes a helical configuration around a single perfusing vessels. The vascularity of the distal bowel is often impaired. UP to 50%may have malrotation .short bowel syndrome is present in nearly 75% of case. Mortality is 54% in this population.

Apple Peel deformity of the small bowel is a variant of Jejunal atresia with high mortality. Forty five percent of these patients can be expected to die, most from anastomotic leaks with sepsis or anastomotic failure with prolonged ileus and malnutrition [3]. This report documents survival of three patients who have atresia near to ligament of teritz, short bowel and large mesenteric gap.

IN this type of atresia bowel blood supply, distal to atresia, is provided from ileocolic or right colic or inferior mesenteric arteries in a retrograde fashion by an anastomotic arcade. For prevention of post operative volvulus in distal segment of intestine in our subjects, we have done a new method of mesoplasty with seromuscular flap of bowel for large mesenteric gap. In this technique, we incised and separated a segment of dilated bowel before anastomosis, with incision and dividing of segment, while preserving of vascular pedicle, with electrocutery denude mucosa of this segment.

This seromuscular flap has been used for large mesenteric defect with fine suturing of the mesenteric edge to this flap. All three patients had a benign postoperative course and made uneventful recovery and no postoperative obstruction was seen.

Based on this experience, I think this method would be a good way in preventing postoperative distal segment volvulus.

پرفوراسیون خود بخودی وراجعه روده در یک شیرخوار ترم : گزارش موردی

Dr Mathew Kripail, Dr Zainab Al balushi, Dr Mohammad.Adellatif

Recurrent Spontaneous Intestinal Perforation in a full term infant: A case report

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Spontaneous isolated intestinal perforation (SIP) typically found in very low birth weight (VLBW) and extremely low birth weight (ELBW) preterm infants is a distinct entity from necrotizing enterocolitis (NEC).

Only few cases have been described in full term infants.these case reports described association with intestinal atresias, anorectal malformations, Hirschprung's disease, segmental absence of the muscularis mucosa in the distal intestine.

We report a case of a full term neonate who was otherwise normal presented with spontaneous intestinal perforation on day 2 of life with no demonstrable cause .The infant had two subsequent episodes of intestinal perforation at six weeks of life requiring surgical intervention with reformation of stoma. All the biopsies revealed nonspecific signs of inflammation and ruled out Necrotizing enterocolitis and Hirschprung's disease.

توده مزانتریک روده

دکتر سهیل بن رضوی

Mesenteric mass of small bowel

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A5 Y/O girl admitted due to lower abdominal pain since 6 months ago. On P/E has no any problem except mild lower abdominal discomfort. Sonography showed a cystic mass in pelvic area. CT scan showed a mass in R.L.Q with Extension to pelvic area.

On Laparotomy a Huge chylous cyst of mesentry of ileum was seen that resected with adjacent bowel completely and anastomosis was done.

Patient was discharged on good condition.

نحوه ترمیم امفلوسل بزرگ با روش کجاوه

دکتر مریم قوامی عادل، دکتر ولی اله محرابی

How to repair giant omphalocele: Case report

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Background: The management of patients with a giant omphalocele remains a difficult problem. Many surgical and nonsurgical methods have been advocated for giant omphalocele management but many of them have difficulties and potential hazards. Here we present an effective surgical method for the management of a large omphalocele easily performs with least complications.

Case presentation: A term girl operated with this method on 9^{th} day of birth. After insertion of two 2 mm pins longitudinally in subfascial position below the defect in both sides of the baby, stay sutures passed around the pins and hanged with elastics from a device like camel litter. The omphalocele sac did not open and covered with a nylon stockings stick to the abdominal wall around the defect which was hanged from the mid portion of the device. After 19 days the liver and intestines reduced easily and the remaining fascial defect repaired with a dual mesh and skin defect covered with a z-plasty. The patient had an uneventful recovery and discharged on 34th days of operation.

Conclusion: This method can be a safe and easy one in large omphalocele repair with minimal complications.

بهبود پی آمد درمان مالروتاسیون روده مطالعه دو بیمارستان کودکان دانشگاه تهران

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Improvement in outcome of children with intestinal malrotation: A study in 2 Children's Hospital

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Introduction and aim: infants with Intestinal mal rotation or rotation abnormalities, addition to the catastrophic risk of midgut volvulus usually associated with others congenital anomalies such as complex congenital heart disease, that required timely, meticulous clinical, radiologic diagnosis and medical management before and after performing a Ladd procedure with or without small bowel resection because known their effects on the survival and post-operative complications. The aid of this study was to review frequency of complications and deaths underwent a Ladd procedure in patients with Intestinal malrotationin recent five years in our two children's hospitals.

Method: The medical records of all patients Intestinal mal rotation who underwent a Ladd procedure In an experience of period of five years of a Team management between 2004 and 2009in two children hospitals centers of TUMS were reviewed. Demographic Clinical symptoms and signs, diagnostic, operative, postoperative complications, and mortality data were recorded.

Results: of sixty -three infants (46 boys and 17 girls) 83.1% underwent an emergency and 15.9% an elective Ladd's procedure after their medical stabilization or palliation of their other anomaly. Range of Age Of these patients at admission were 1day to2months

The median age of patients at the time of the operation was 5.7days .the most common clinical symptoms was bilious vomiting in 88.9% of patients and median stay days of hospitalization was 6.4 days (ranged between3and15days).

Range of Age Of these patients for evaluation for complications was between 2.3 and 6.04 months after the operation. Developed postoperative intestinal obstruction due to adhesion bands: lysis of adhesions was occurred in 5.9%patient; 7, 9%patient required another operation for a recurrent midgut volvulus after an incomplete initial Ladd procedure. The 6/3 deaths, after the surgery, occurred as sequelae of the underlying cardiac anomaly. Length of follow-up ranged from 2.3 months to 5 years.

Conclusions: our recent 5 years' experience suggests by a team management, preoperative support with timely performing Ladd procedure of patients with malrotation has an acceptably low morbidity and. Allow risk of 12.2% of postoperative bowel obstruction comparing to significant risk of mid gut volvulus.

دوپلیکاسیون متعدد روده یک گزارش موردی وبررسی متون

دکتر دیانا دیاز

Multiple Intestinal Duplication: A case report and brief entity review

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Introduction: Intestinal duplications are very uncommon congenital malformations located in the mesenteric edge of the small bowel, particularly in the ileum. Over 60% of the patients become symptomatic during the first year of life whereas the remainder demonstrates symptoms at school age or adulthood. The wide spectrum of symptoms and unspecific signs frequently simulate other diseases. Gastrointestinal hemorrhage is the most noteworthy complication, which can cause severe anemia and shock.

Aim: In this presentation the intestinal duplication is analyzed and the importance of considering it as a differential diagnosis in any kind of bleeding of the digestive tube is discussed.

Methods: In the case we describe an 8 month-old boy who experienced massive intestinal hemorrhage and anemic syndrome. In this patient, hemoglobin levels rose up to 5 g/dL necessitating hemotransfusion. Multiple diagnosis and examinations were carried out until finally the gammagram with Tc99m disclosed an ectopic gastric mucosa. Using a laparotomy three intestinal duplications were found in the mid jejunum that were resolved by means of bowel resection and anastomosis. The post-op course was uneventful and the patient was discharged at the fourth day.

Conclusion Pediatric patients with unexplained gastrointestinal symptoms must be diagnosed for the possibility of alimentary tract duplication, even though it rarely occurs. With proper treatment, children born with enteric duplications should do well and have excellent long-term outcomes and quality of life.

پی آمد آترزی قسمت فوقانی دستگاه گوارش در یک مرکز اطفال

دکتر مریم قوامی عادل ، دکتر سید مجتبی موسوی خوشدل، دکترسید جواد نصیری، دکتر فریبا جهانگیری، دکتر پیروز فرهود، دکتر صلاح الدین دلشاد

Overview of outcomes in upper gastrointestinal atresia in a pediatric center

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Background-Intestinal obstruction occurs in approximately 1 per 500-1000 live births and about 50% is due to atresia. Despite improvements in care and survival, intestinal atresias are associated with prolonged hospitalization and occasionally mortality. Although each type of atresia is distinct, there are obstacles predicting the outcome. This study performed to identify factors that may predict the outcome.

Material & Method- Records of patients with small intestinal atresia were analyzed retrospectively (2006-2011) for location, demographics, prenatal diagnosis, diagnostic evaluations, birth weight, associated anomalies, cost, beginning the oral feeding, and mortality. SPSS 18 used for data analysis.

Results- A total of 40 infants with 20 males and 20 females were evaluated including 28 with duodenal, 11 with jejunal/ileal, and 1 with pyloric atresias. Associated congenital anomalies identified in 19 patients with 6 Down syndrome. There were 22 preterm infants with mean birth weight 2365.38 grams (1700-3700). Overall mortality was 22.5%. Using Binary logistic regression, there was no correlation with sex (P=0.7), birth weight (P=0.49), associated anomalies (p=0.46), Down syndrome (0.63), type of atresia (P=0.35), presence of cardiac anomaly (p=0.17) and mortality, but mortality was significantly higher in preterm infants (P=0.04). Also there was no correlation between the duration of hospitalization and birth weight (P= 0.74), the site of atresia (P=0.99), associated anomaly (p=0.37), interval between admission and operation (p=0.69) and gestational age (p=0.05).Beginning the entral feeding had no correlation with weight (P=0.31), associated anomalies (p=0.5), site of atresia (p=0.33) and gestational age (p=0.17)

مقایسه دوره بیدردی "بیوپیوکائین"،"بیوپیوکائین- میدازولام "و بیوپیوکائین- کتامین" برای بی حسی کودال در عمل های جراحی پائین شکم در کودکان

دکتر ابراهیم اسپهبدی، دکتر احسان نجمی

Comparison of analgesic duration of Bupivacaine and Bupivacaine - Midazolame and bupivacaine –ketamine for Caudal Anesthesia in Children with lower abdomen surgery

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Background: Caudal anesthesia is one of the most popular regional blocks in children (1). This technique is usually performed after an inhaled or IV induction and is a useful adjunct during general anesthesia and for providing postoperative analgesia after genital, lower abdominal, and lower limb operations (2). We performed a prospective randomized double-blinded study to test Bupivacaine alone or in combination with preservative-free S (+)-Ketamine and Midazolame for intra- and postoperative caudal blockade in pediatric surgery over a 24-h period.

Methods: The study protocol was accepted by our department review and ethical committee. We obtained written informed parental consent to perform this study. 60 children (1–3yrs) were divided into 3 groups of 20 in each scheduled for lower abdomen surgery were caudally injected with Bupivacaine 0/25% 1cc/kg (Group A), Bupivacaine 0/25% 1cc/kg +Ketamine-s 0/5cc/kg(group B) and Bupivacaine 0/25% 1cc/kg+ Midazolame 50mic/kg (group C) during Sevoflurane anesthesia

Intraoperative monitoring included heart rate, blood pressure, and pulse oximetry; postoperative monitoring included a pain discomfort scale and a sedation score. No additional analgesic drugs were required during surgery.

Results: The observed data showed that the mean duration of postoperative analgesia was 19 hours in group C, 14.5 hours in group B and 8 hours in group A. nausea; vomiting and psycho logic reaction was higher in group B.

Conclusion: We conclude that the combination of S (+)-Ketamine 0/5 mg/kg or Midazolam 50mic/kg with Bupivacaine 0/25% 1 cc/kg for caudal blockade in children provides excellent analgesia without significant side effects over a 24-h period.

تنگی آناستوموز کولوآنال شایع ترین عارضه جراحی ترانس آنال پولترو بیماری هیرشپرونگ

دکتر محسن روزرخ، دکتر احمد خالق نژاد طبری، دکتر لیلی مهاجرزاده، دکتر حیدری، دکتر مولایی

Coloanal Anastomosis Stricture as a most common compilication of Trans anal pullthrough in patients with Hirshsproung disease

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Background: Hirschsprung's disease (HD) is a relatively common congenital disease that could be suspected by clinical symptoms, abdominal plain X-ray, and finally diagnosed by rectal biopsy. In 80% cases, rectosigmoid junction is involved. Recently, one-stage transanal pull-through (TAPT) procedure has been popular and may have several complications.

Method and patients: During a 4-year period, 86 infants (28 girls, 58 boys) with mean age 8 days (3-33) and clinically suspected to HD were admitted in our center. HD was proved by rectal biopsy. All patients after full bowel preparation and rectal washout were candidates for TAPT operation. A Swenson-like procedure was performed and the anastomosis was done between the well blood supply ganglionic colon and the rectum at 1 cm above dentate line. Interrupted suture with 5-0 Vicryl was used. Nelaton tube (12 F) inserted in the pelvis via transprineal for drainage of blood or collection. From February 2008 in 30 cases, prophylactic Hegar dilatation was performed 2 weeks after operation.

Results: Anal stricture in 12 cases (14%) was treated by anal dilation in 10 cases and 2 cases corrected by surgical management. Entrocolitis in 4 cases (5%) was treated by medical management. In two cases, retrocolic abscess had spontaneous drainage via tube drain. There was no anastomotic stricture after starting prophylactic anal bouginage.

Conclusion: TAPT has many advantages, low complications and the results are excellent. It seems the most common complication is anastomotic stricture that responds well to prophylactic bouginage. We recommend prophylactic anal bouginage with Hegar probe at 2 weeks after operation. Long-term follow-up is needed to evaluate the outcomes of our operations.

کولکتومی توتال و آناستوموز ایلئورکتال همراه با مایاتومی انورکتال روش جدید درمان هیرشپرونگ توتال کولونیک و اختلالات حرکتی روده

دکتر احمد خالق نژاد طبری، دکتر حمید فروتن، دکتر عباس بنانی، دکتر لیلی مهاجرزاده، دکتر محسن روزرخ، دکتر علیرضا میرشمیرانی

Total Colectomy and Ileorectal Anastomosts with Anorectal Myotomy: a new procedurefor treatment of total colonic aganglionosis and gastrointestinal dysmotility

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Introductions and Aims: We have performed State pull-through as ileoproctostomy with long posterior myotomy in total colonic aganglionosis and sever dysmotility disorders.

Methods: 13cases, 10 total colonic aganglionosis, 1intestinal neuronal dysplasia (IND) and 2 chronic intestinal pseudo obstruction syndrome (CIP)) from1992 to2012 underwent total colectomy and resection of part of involved small intestine and ileorctal anastomosis in one layer with 4/0 Vicryl with long posterior rectal myotomy. All patients had barium enema and rectal biopsy.

Leveling ileostomy was done in 12 cases of which one had distal jejunostomy.2 of 13 children, proximal diverting loop ileostomy had been established. 3cases, 2 weeks after initial operation, myotomy from anus, performed.

Results: 13 female patients at the age of 6 months to years. Rectal biopsy of10 Patients reported no ganglion cell of which one had extended aganglionosis to distal jejunum, one had IND and two had ganglionic bowel with clinical presentation of CIP.

Follow up time was 6months to 10year.There were no significant complication in this group of patients except episodes of diarrhea and severe dehydration that need hospitalization and hydration. .All have acceptable bowel function following operation (2-6 times a day).Now, 5 children of 13patients are above the age of toilet training have voluntary bowel movement with little or no medication (Leopromid).

Conclusion: State pull-through is recommended in all cases of total colonic aganglionosis and severe dysmotility problems of colon. This technique is less difficult to perform, and avoid the complications and disadvantages of removal of the rectum and has satisfactory results.

درمان بیماری هیر شپرونگ بروش ترانس آنال در نوزادان مطالعه ۴ ساله

دکتر بهار اشجعی، دکتر جواد احمدی

Trans anal pull through in neonates with Hirshsproung disease: 4years experience

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Background: Hirshcsprung's disease or absent of ganglion cells in bowel is a congenital disease that is cured by resection of aganglionic segment of the bowel.

In the past decades a stoma was placed in distal of ganglionic segment before pull through procedure and at least the operation was done in two stages. Today trance anal pull through is done in one stage without any stoma placement.

Material and methods: in this study we performed a rectal biopsy for any neonate who was admitted in our hospital and suffering from delaying of me conium passage for at least 48 hours without any predisposing factor and the barium enema were suspicious of HD. We performed trance anal pull through in cases that ganglion cells were not seen frozen section pathologic evaluation. In patients whose rectal biopsy had ganglion the operation were finished without any other procedures.

Results: we recognized 23 patients with HD by this method in 4 years and all of them underwent one stage trance anal pull through. The hospital stay was lower than the traditional methods of pull trough and the main complication was barium trance location in retroperitoneum in one case who was underwent barium study before operation. We had not other serious complication in other cases. Furthermore in all cases the cosmetic results were better than traditional methods.

Conclusion: one stage trance anal pull through can be a good replacement for the other methods of pull trough in patients with irschsprung's diseses.

پوستر شماره1

بررسی شیوع آترزی مری در سال 1389 در مرکز آموزشی درمانی کودکان تبریز

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An investigation into the prevalence of esophageal atresia in 1389 in Children Educational-Health Center of Tabriz

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Introduction: esophageal atresia is one of the anomalies of digestive system that after imperforate anus is the most common digestive disorder. Esophageal atresia is categorized in four, A, B, C, and D groups. This anomaly is equally observed in both genders. Symptoms such as frothy saliva, watery mouth, chocking, cough, sneeze and cyanosis are observed. For diagnosis of the disease a tube is sent to the esophagus through the nose and if the tube is blocked somewhere then the place of the esophageal atresia is probably found.

Procedure: this study is a research that aims to specify the prevalence rate of esophageal atresia. The data are collected using the patient files who were hospitalized in 1389. In this research, 40 cases from 1201 hospitalized patients were suffering from esophageal atresia and entered our statistical population. SPSS software was used in order to analyze the data.

Results: the results from the study showed that in 1389 the prevalence rate of esophageal atresia was 3.3% and from each 40 cases, 22 were children; 75% of the patients were suffering from type C esophageal atresia; 55% were treated by mechanical ventilation and 27.5% of the patients died.; 40% of the patients suffered from heart disease and the disease was observed equally in both genders.

Discussion: since the quick diagnosis is in first priority and without rapid diagnosis and operation of modifying surgery the disease will rapidly lead to death it is much better to diagnose the disease in a delivery room. The survival rate is high in case the patient is only suffering from the disease itself. The most mortality is seen when the disease is associated with other deadly anomalies. In this investigation we observed 11 deaths in 40 cases of the disease and from these 11 cases, 6 cases also had heart disease (VSD, PDA, ASD, CHD, pulmonary atresia hypoplasia of the left ventricular and etc.), 2 cases were associated with imperforate anus, 1 case had Down syndrome and 1 case died before surgery.