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Pre- and Postnatal Surgery, Most Common Conditions, Diagnosis and Treatment

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Abstract

The authors make a general but relevant description of the most common surgical problems occurring both in the human being that develops in the uterus and in the one that has already been born.

Keywords: prenatal surgery, postnatal surgery

1. Introduction

Congenital malformations, apart from the achievements in the prenatal care of women with high-risk pregnancies, are still a subject of great importance in our country. As Mexico is a developing country, due to various factors, an expeditious attention to newborns with malformations has not been achieved. An additional issue that affects the treatment of these children is the alarming increase in the prevalence of structural anomalies. Health statistics indicate that the birth rate is among the world's highest, which means that sometimes the state is incapable of providing the required assistance. Socioeconomic and educational factors are among those that have contributed to the increase in congenital malformations in the past 10 years, and the desired rate of recovery of these children has not been reached. Because of the above situation, specialists have had to refine their skills in elaborating the clinical diagnosis and instituting the appropriate therapeutic measures. In Mexico, neonatal surgery has become a specialty recognized by general pediatric surgery and by the Autonomous National University (UNAM).

2. Prenatal

Maternal-fetal medicine and intrauterine intervention in defects of the fetus, or unborn child, emerged and has evolved as a result of the technical advancements in medicine. The scope of these specialties is encouraging and promises advanced interventions with improved results. The refinement of ultrasound has made a great contribution to the field [1]. The use of ultrasound to visualize the fetus in its environment created a great interest to be able to identify malformations that could be corrected through intrauterine intervention. The most important antecedents of fetal surgery date back to the early 1960s, when Liley devised a technique for fetal transfusion [2]. In 1965, Adamson reported the first fetal surgery in humans, using a hysterotomy [3]. In this research line, experiments were carried out on lamb and monkey fetuses to perfect instruments and techniques, evaluate the results in these models and study their feasibility and reproducibility in humans.

The modern history of fetal surgery began at the University of California, San Francisco, with Michael Harrison. He first conceived his interest in intrauterine intervention for the treatment of congenital diaphragmatic hernia when he was an intern at Massachusetts General Hospital. Harrison's work continues today and is a valuable resource in the field [1–3]. In 1982, Harrison noted that diaphragmatic hernia, congenital hydronephrosis and hydrocephalus were defects which could be susceptible to prenatal treatment, as they were simple structural defects that prevented the normal fetal development of the structures involved. In that same year, the first open fetal surgery was performed at 21 weeks of gestation on a fetus with congenital hydronephrosis, which consisted of performing a hysterotomy to expose the lower abdomen of the fetus and place a double pigtail catheter to communicate the bladder with the amniotic cavity to promote the free flow of urine, thus relieving the obstruction causing the hydronephrosis [4]. The intervention was successful, and pregnancy continued for 14 weeks after intervention; however, at birth, the kidneys already showed irreversible damage, and it was clear that intervention had to be at an earlier stage of intrauterine life [5].

Neonatal surgery was not well known as a separate specialty of general pediatric surgery but has a history dating back to 1955 when P. Rickham, a pediatric surgeon from Switzerland, started a newborn surgery program in the city of Liverpool. Rickham's program began the systematization of neonatal care, especially for congenital anomalies. It was to be hoped that, having established a program of neonatal surgery, it would be recognized as a specialty. In spite of his prestige and worldwide fame, the specialty was not yet established. He published two editions of his classic book *Neonatal Surgery*, where in addition to the classically studied congenital defects, he narrated his experience operating on children with neurological conditions such as congenital hydrocephalus associated, or not, with pituitary-spinal malformations and musculoskeletal disorders such as club foot.

Neonatal surgery in the last third of the twentieth century has made remarkable advances in diagnostic resources. Postoperative management development has been especially important, with the implementation of ventilators and intravenous nutrition. In Mexico (specifically in the unit where the authors work), after years of managing hundreds of malformed newborns,

a successful postgraduate project in neonatal surgery has been developed at the Universidad Nacional Autónoma de México, which is already being replicated in other institutions in Mexico and other Latin American countries.

2.1. Obstructive uropathy

Congenital hydronephrosis was the first condition to attract attention within the scope of prenatal interventions, especially in bladder outlet obstructions due to urethral valves [4]. Congenital obstruction of the lower urinary tract (bladder neck) comprises a set of conditions of which atresias and posterior urethral valve obstructions are representative. With an approximate incidence of 2.2 per 10,000 live births, it results in oligohydramnios, pulmonary hypoplasia and irreparable renal damage, with 45% mortality. The role of prenatal obstetric ultrasound is crucial as the defect can be detected in 85% of the cases. Ultrasonographic features suggestive of lower urinary tract obstruction are an enlarged fetal bladder and dilated proximal urethra with or without associated hydronephrosis. It has been observed that when the anomaly is present before 25 weeks of gestation and secondary oligohydramnios prevails for more than 14 days, this malformation is associated with neonatal mortality > 90%. The canalicular phase of lung development, crucial to the development of the human lung, occurs during 14–25 weeks of gestation. Morris et al. demonstrated that prenatal intervention to treat bladder obstruction by hydronephrosis using percutaneous or open bladder shunt by a vesico-amniotic shunt increases overall survival in neonates affected by this abnormality, however, the studies published in this regard are few [6].

2.2. Sacrococcygeal teratoma

Sacrococcygeal teratoma is the most common tumor in the newborn. This neoplasm arises between the base of the spine and the rectum and protrudes from inside of the pelvis outwards. The tumor is more frequent in females than in males (3:1), with an incidence of 1 per 40,000 births. The presentation may be grossly frank and is seen at birth. Type 1 has the best prognosis, as the tumor is predominantly external with a small pre-sacral component (**Figure 1**). The tumor corresponding to type 4 is invasive, deforming the pelvis and part of the abdomen. The risk of malignancy depends on two factors: the site and extent of the tumor and age at the time of diagnosis. The prenatal appearance of the sacrococcygeal teratoma on ultrasound is of a mixed solid lesion with a cystic component that arises from the sacrum, with hyperechogenic zones corresponding to calcifications. Recently, ultrafast fetal nuclear magnetic resonance imaging has been used to assess the vascular component of these neoplasms, as there is risk of fetal intrauterine hemorrhage with secondary anemia, hemodynamic decompensation due to the formation of arteriovenous fistulas, hydramnios, hydrops fetalis, placentomegaly and eventually, in severe cases, intrauterine death. The effect of the mass of a sacrococcygeal teratoma has been related to uterine distension, dystocia and tumor rupture at birth, as well as causing mirror syndrome and severe hypertensive states.

The hemodynamic effect of the coccygeal sacrum tumor can be evaluated with echocardiographic study, measuring the relationship between ventricular outflow and the diameter of the vena cava, descending aorta and umbilical vein. A poor prognosis is related to



Figure 1. A giant sacrococcygeal teratoma in a newborn patient.

disproportionate tumor size (more than 150 cm per week is a very poor prognosis), high vascularity, predominantly solid component, placentomegaly, heart failure and concomitant maternal complications. The main objective of prenatal care is to identify the most reliable predictor of poor prognosis in fetuses with sacrococcygeal teratoma. The gestational age at the time of decompensation will determine the type of intervention; if it occurs after the 27th or 28th week of gestation, an emergency cesarean section or EXIT procedure (ex utero intrapartum treatment) will be chosen. If the hemodynamic imbalance is identified before week 27 in the cases of teratoma classes 1 and 2, open maternal-fetal surgery and partial resection of the tumor followed by definitive surgical intervention at birth should be chosen. There are other procedures that consist of disrupting tumor vascularity with dissimilar results, such as thermoregulation, radiofrequency ablation, alcohol sclerosis or laser ablation. The complications or side effects of these methods can result in extensive pelvic muscle damage, hemorrhage, nerve injury and hip dislocation [7].

2.3. Myelomeningocele

Within the congenital anomalies of the central nervous system, myelomeningocele is the most common variety of spina bifida and consists of the extrusion of the spinal tissue into a sac (meninges) occupied by cerebrospinal fluid. Its frequency is about 3.4 per 10,000 live births. Mortality is 10%, and survivors show varying degrees of disability depending on the height of the neural damage, including paralysis of the lower extremities and bladder and bowel dysfunction. In addition to exhibiting the anatomical neurological abnormalities characteristic of

Arnold-Chiari malformation. This anomaly is a generally congenital disease, consisting of an anatomical alteration of the base of the skull, which produces herniation of the cerebellum and brainstem through the foramen magnum to the cervical canal. This anomaly ultimately causes abnormal circulation of cerebrospinal fluid, culminating with hydrocephalus, central apnea, stridor and swallowing disorders. The embryonic pathophysiology of the defect is due to the combination of two strands or the two-hit model. The first effect consists of failure in the formation and closure of the neural tube, with an open defect, which exposes the neural elements to amniotic fluid (the second effect). It is important to note that in experimental models with fetal lambs, where an open neural defect was created and immediately surgically covered, effects on the lower extremities and on urinary continence and bowel movements were not observed. On the other hand, fetuses of lambs who underwent surgical repair of the myelomeningocele after 4 weeks of having experimentally created the defect showed good outcome after fetal intervention, urinary continence was maintained and preserved the neuromuscular function of their limbs, Which supports the hypothesis that prenatal intervention prevents neurological and associated confusion. Adzick in 2011 in a randomized clinical trial [8].

2.4. Pulmonary cystic adenomatoid malformation

Congenital pulmonary malformations have been divided for their study into those related to the bronchial tree: agenesis, aplasia and pulmonary hypoplasia and those of pulmonary parenchyma: congenital lung cystic adenomatoid disease, pulmonary sequestration, lobar emphysema and bronchogenic cyst. The high definition and quality provided by current sonographic studies have achieved better prenatal identification of the fetal lung lesions in the majority of cases detected on routine ultrasound between 18 and 20 weeks of gestation. These malformations have an incidence from 1 per 10,000 to 1 per 35,000 pregnancies. Sonographic study provides information on variables such as volume, location, arterial blood supply and venous drainage and consistency of the adenomatoid malformation, which can vary widely from solid (microcystic) to frankly cystic (macrocytic) [7].

Adenomatoid malformation, consisting of small cysts, generally grows unpredictably and causes compression on surrounding structures, as well as an effect of mass on the mediastinum, esophagus and lungs, causing pulmonary hypoplasia, obstruction of the vena cava, cardiac insufficiency, hydrops fetalis and polyhydramnios. As one method to identify serious cases, several investigators have proposed predictive indices and measures such as the cystic volume ratio (CVR), obtained by dividing the volume of the cystic lesion (length X width X height X 0.52) by the circumference of the head. In a retrospective study by Crombleholme, it was concluded that a CVR greater than 1.6 was associated with a 75% chance of developing hydrops fetalis, which justified prenatal intervention [7, 9]. In cases with a better prognosis, EXIT therapy is recommended, with resection of the mass before birth. Postnatal stabilization and thoracotomy are also acceptable. Macrocytic lesions may be prenatally decompressed by thoracentesis with a single needle or with drainage guided by amniotic thoracic ultrasound. At this time, microcystic lesions are not candidates for drainage and will require fetal surgery [10].

3. Postnatal

3.1. Esophageal atresia

Esophageal atresia is one of the most common congenital malformations and perhaps the most classic of structural abnormalities that exclusively involve the pediatric surgeon, as only a specialist can resolve the problem. Other specialists such as the urologist usually operate on an undescended testicle and a general surgeon on a pyloric stenosis, but even a thoracic surgeon would not attempt to correct an esophageal atresia. The frequency of the anomaly is one case per 3000 to 5000 births, predominately in males, and the prevalence may depend on the region or country being analyzed. How esophageal atresia occurs is unknown, but there are many theories, none of which has withstood the test of time. Esophageal atresia is usually accompanied by other defects, so acrostics are formed to list the components such as VACTERL, for vertebral, anorectal, cardiac, tracheo-esophageal, esophageal, renal and limb anomalies [11]. At times, it is sporadically associated with other defects of the digestive tract, such as duodenal atresia. In Mexico, the frequency measured in cases per year is between 12 and 16 cases a year in regional and specialty hospitals. The Moctezuma Pediatric Hospital, a specialty hospital of the Secretary of Health of Mexico City, sees the most cases per year [12].

3.1.1. *Diagnosis*

No specific biological teratogen is known, but the anomaly may be produced by the experimental administration of Adriamycin. More often than not, it is a sporadic malformation that does not obey genetic or hereditary patterns, although it is usually accompanied by trisomies such as 13 and 18 or those associated, without the atresia being the axis of clinical expression. It usually occurs in the products of conception of young women. Polyhydramnios can be found during the prenatal visit, which should be the most important warning sign [13, 14]. In pediatric surgery, it is said that sialorrhea in the newborn child is equivalent to an esophageal atresia until proven otherwise. If this is accompanied by cyanosis and asphyxia during feeding, the diagnosis is confirmed. Although it should be noted that it is currently the obligation of the obstetrician in the delivery room to make the clinical diagnosis of suspicion. In other words, waiting for the child to choke on ingested milk signifies malpractice and tardiness to integrate the diagnostic impression.

The routine introduction of a nasogastric catheter into the stomach is a maneuver that should be performed in every delivery room. It is the most effective, advisable and efficient way for a child with esophageal atresia to achieve a good condition without additional risk during intervention. With the diagnosis confirmed, the next step is to inject 0.5 ml of water-soluble contrast medium for radiological study (**Figure 2**), which, in addition to highlighting the height of the atresia, will demonstrate a proximal fistula if it exists and for which a surgical procedure would be indicated [13]. When the defect is detected, a probe must be left in the proximal pouch so the saliva is prevented from spilling over into the respiratory tract. The tube should be constructed as a trap so as to provide continuous suction without pinching the esophageal mucosa and avoid blockage. This is achieved by introducing a thin probe through the counter-aperture in the middle third of a larger probe. The thicker probe should be very close to the

bottom of the pouch, and the thinner probe will serve to continuously instill saline solution to keep the system efficient.

From the clinical point of view, esophageal atresia has been divided into four varieties and classified into letters or numbers. The system with letters is the most used: type A, two blind pouches, but no fistula; type B, two segments with the upper one connected to the airway; type C, two segments with the distal esophagus in the airway; and type D, both segments that contain a fistulous path. Radiological studies, in addition to revealing the aforementioned findings, may show a cardiac silhouette that suggests right aortic arch cardiopathy, as the presence of some skeletal malformations such as hemi-vertebra or spina bifida can be detected. The prognosis may be definitive when an area of pulmonary consolidation is identified. The presence of air in the digestive tract indicates the existence of the most frequent variety, an atresia type C or D. The absence of air in the digestive tract identifies variety A or B. Once the diagnosis has been confirmed, studies to reduce the risk to the child should be made. Thus, when a condition that has a high risk for esophageal atresia is detected, such as hydrocephalus or obstruction of the urinary tract, it should be given priority [15].

3.1.2. Treatment

The management of esophageal atresia requires zeal and closeness to the patient as with few other anomalies. In no other condition is it so important for the pediatric surgeon to be aware that the suction device works and the child's vital signs and laboratory values have been restored. Antibiotics are essential. In ideal conditions, an end-to-end esophageal anastomosis

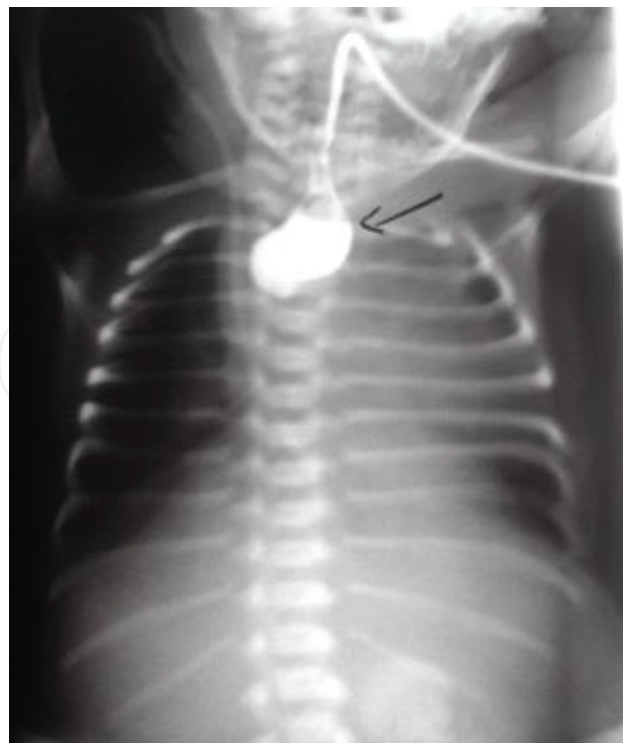


Figure 2. Simple thoracic X-ray, revealing the fundus of the proximal sac contrasted with a water-soluble medium. This is an esophageal atresia type A.

should be performed by right thoracotomy, regardless of whether it is in one or more anatomical planes. It is important that it is done with fine, inert material and the placement of the stitches is firm and their number and location are correct. Ideally, it should be done outside the pleural space, but if it is not feasible, a pleural tube should be placed and removed on the day of the esophagogram to prove that there is no dehiscence and/or refistulization. In approximately a week, they may begin oral feeding; meanwhile, they must be maintained intravenously.

Prognostic criteria can identify those who are included in the favorable group and those who will survive. The best-known criteria have to do with the weight of the product of conception, the presence of other malformations and if bronchial aspiration pneumonia is present. Based on these criteria, those classified as risk type A are those weighing more than 2500 grams, lacking significant malformations and without pneumonia, and more than 90% of these cases survive. Mortality is 50% or more in the opposite, high-risk conditions. Complications, which can be classified as immediate and mediate, are frequent. In the former, it is the dehiscence of the anastomosis, which always has a reserved prognosis, and during late onset, it is the pathological gastro-esophageal reflux [15].

3.2. Duodenal atresia

Atresia of the duodenum is the congenital obliteration of any portion of the small intestine, which lies between the most distal pylorus and the most proximal part of the ligament of Treitz. This is a very common malformation and almost a permanent occupant of neonatal intensive care units, as one case appears for every three to five thousand births [16]. It almost always appears as a sporadic condition but may be part of some chromosomopathies such as trisomy 21. It has been assumed that it appears between 5 and 12 weeks of gestation. The most accepted theory is that of Tandler. Known as recanalization, it consists of the appearance of the concentric histolysis phenomena due to the invasion of vacuoles into the solid cord, an essential characteristic of the duodenum in the embryonic stage, to the degree that the invasion converts the cord in its definitive form, into a tube [17]. Other theories, such as ischemia may have a basis, especially in segment atresia accompanied by a loss of duodenal continuity with or without intermediate fibrous cord.

Anatomically, duodenal atresia has been divided into two forms: the proximal and distal to the ampulla of Vater and divided into three forms clinically—when it appears as a septum or as “wind sock,” when there is an interruption of continuity and when these segments are joined by a thin fibrous cord. Regarding the wind sock variety, it should be emphasized that duodenal blockage does not appear at the transition site of the dilated proximal segment and the thin distal segment but that the membrane always emerges a few millimeters above the narrow area.

3.2.1. *Diagnosis*

Duodenal atresia usually appears early in pregnancy, accompanied by polyhydramnios. When performing sonographic screenings in prenatal consultation, the presence of polyhydramnios should alert the specialist to look for data suggesting blockage of the digestive tract, such as atresia of the duodenum or of other distal structures. Once the product has been born, the correlation can be made by gastric lavage with a catheter as a routine measure and obtaining 20–25

ml of transparent or biliary fluid, which indirectly evidences alterations in the dynamics of the movement of amniotic fluid. If the mongoloid facies resulting from trisomy is added as relevant data, the suspicion of a duodenal block is strengthened. On inspection, there is an increase in volume of the upper abdomen, the skin is taut and shiny and there is early vomiting. Depending on the level of the blockage, the expelled liquid will be yellow, thick and abundant if the blockage is below the ampulla of Vater and the vomitus will be transparent if the obstruction is proximal to the main bile duct outlet. Due to alterations in the enterohepatic circulation, this anomaly is almost always accompanied by jaundice, which disappears as soon as the problem is resolved through surgical intervention. If several days elapse before identifying the anomaly, from the clinical viewpoint, there are usually hydroelectrolytic alterations, exacerbation of jaundice and signs of a systemic inflammatory response. Regarding radiology, simple radiography of the abdomen in an upright position is sufficient, as air is the best contrast medium in those cases. When children have a tube inserted in the stomach, 15–20 ml of air should be injected at the time of the study in order to maintain the contrast. Observing the image of the “double bubble” (**Figure 3**), it is a certainty that the duodenum is totally or partially blocked, and there is a presupposition of the existence of defects such as annular pancreas, malrotation, presence of a narrow angle mesenteric artery and the preduodenal portal vein. However, the presence of the facies, umbilical hernia and macroglossia are usually diagnostically determinant factors so that before the surgical intervention, with a good degree of certainty, it is assumed that the baby suffers from duodenal atresia. No endoscopy or introduction of liquid contrast material is necessary.

3.2.2. *Treatment*

In our country, this type of patient usually arrives in a serious condition. Vomiting is early, and in less than 24 h, there is an important loss of fluids and electrolytes, so the patient must be immediately transferred to an intensive care unit, where a gastric tube must be installed to allow the exit of all the liquid that is lodged in the stomach in utero, a quantity much superior to the normal gastric capacity. Antibiotics should always be used, as there is often respiratory infection aggravated by microaspirations. Once the gastric tube has been installed and the patients' condition improved, the patient must be transferred to the operating room where, depending on the findings, the intervention of choice will be instituted. If confronted with an atresia without loss of duodenal continuity (variety I), a slight concentric depression should be located on the surface of the duodenum. That indicates the point where the obstructive diaphragm is inserted. Once this reference is placed, a horizontal incision is made on the depression, the septum is removed, the mucosa is sutured with a continuous line of absorbable fine material and the duodenal wall is repaired vertically. This method is known as the Heinecke-Mikulicz technique. If, on the contrary, atresia shows separation of segments, then it must be resolved through two types of access: through a minimally invasive procedure consisting of the introduction of very thin laparoscopy tubes and laparotomy [17]. In both methods, the surgical technique will be the duodenal-duodenal diamond-shaped anastomosis, proposed by Kimura et al. [18], which consists of joining both blind segments, making a horizontal incision in the proximal and a vertical one in the distal so that when anastomosis is terminated, intercommunication is wide enough for the intestinal flow.



Figure 3. Simple abdomino-thoracic radiological study showing great gastric dilation and obstruction of the second portion of the duodenum.

The original report of the diamond anastomosis technique says that the oral feeding route in the postoperative period could be initiated in the first 5–7 days; however, in our experience [19], the waiting period for tolerance of fluid intake can be extended up to 4 weeks, which means that those children must undergo intravenous nutrition for at least 15 days. We believe that duodenal dilatation resulting from the receipt of large amounts of fluid results in alterations in emptying movements, which is corrected spontaneously but later in postnatal life. An additional problem is that these infants suffer from recurrent nosocomial infections due to their prolonged hospital stay and the use of a catheter, which makes their recovery more difficult. Management also includes a gastric probe that protects the anastomosis, the use of antimicrobials and analgesics.

3.3. Jejunoileal atresia

The small intestine is a complex and vital organ. Jejunoileal atresia accounts for almost one-third of all cases of intestinal obstruction in the neonatal period.

The pathogenesis of jejunoileal atresia is still controversial. In the middle of the last century, it was proposed that the atresia was due to a failure in recanalization; however, Louw and Barnard, through experimental studies, showed that this anomaly occurs as a consequence of an ischemic phenomenon that culminates with segments of mesenteric ischemia and in atresia. Subsequently it was shown that abdominal catastrophic events in utero were responsible and that the most common conditions were invagination, perforation, volvulus, strangulation by an internal hernia or thromboembolism. The spectrum of atresia depends on the extent, severity and duration of the ischemia, since the mucosa and submucosa are more susceptible. An incidence from 1 to 5 per 10,000 live births has been estimated. It affects both sexes equally. It affects both the jejunum and the ileum similarly. Compared to duodenal atresia, associated anomalies are less frequent. Multiple intestinal atresias can be an autosomal recessive disorder, more commonly seen in combination with some degree of immunodeficiency and in Canadians. Mutations have been observed in the TTC7A protein, important for the development and function of the intestinal epithelium. This mutation has also been associated with early presentation of inflammatory bowel disease [20].

Based on their anatomical characteristics, four types of atresias have been described. Type I is an intramural membrane of mucosa and submucosa that is continued with a cord, with no mesenteric defect. In Type II, there is intestinal discontinuity but no mesenteric defect. Type III has two subtypes; in subtype IIIA, continuity of the intestine is interrupted and there is also a V-shaped defect of the mesentery. In subtype IIIB, there is a lack of continuity of the intestine and it is wrapped around the superior mesenteric artery, giving the appearance of a Christmas tree or apple peel. Type IV consists of a segment with multiple atresias that resemble a string of sausages. We have designated a type V, which consists of a combination of atresias types I, II and IIIA present in the same intestinal segment [21].

3.3.1. *Diagnosis*

The typical presentation is that of a newborn on the first or second day of life with biliary vomiting, a history of polyhydramnios and abdominal distension, which will be more severe the more distal the obstruction. The most serious form of presentation is intestinal perforation. The diagnosis can be made prenatally through the use of ultrasonography. Findings suggesting atresia, in nearly one-third of cases, include dilated bowel and polyhydramnios. If there is strong suspicion, fetal magnetic resonance usually determines the diagnosis. Once the product of conception is born and suspected, simple vertical and horizontal x-rays of the abdomen should be taken. When a jejunal atresia is present, X-ray will show multiple hydroaerial levels (**Figure 4**) and the “triple bubble” sign. The presence of calcifications suggests the catastrophe and may be present in about 12% of cases. In that scenario, confirmation of atresia is obtained by colonic enema, when a microcolon is observed [21, 22].

3.3.2. *Treatment*

Once the diagnosis has been made, a gastric tube is placed to empty the proximal digestive tract, the hydroelectrolytic alterations are corrected, antibiotic management is added and a central venous catheter is installed. The operation can be performed by an open or a laparoscopic

approach, in order to resect the proximal atresic bulbous segment and perform a primary anastomosis with the distal segment. A precautionary transoperative routine measure is the instillation of saline solution to the distal intestine in order to rule out the presence of other obstacles [22, 23].

Mortality is related to prematurity, associated anomalies, infections and short bowel syndrome. Postoperative complications that may occur are leakage, stenosis of the anastomosis site and short bowel, if there was extensive bowel resection. Food intolerance may be a mediate complication. Success is rated by the time of initiation of enteral feeding, postsurgical complications and the duration of feeding with total parenteral nutrition. The prognosis of neonates with jejunoileal atresia is very good, with a survival rate greater than 90%.

3.4. Necrotizing enterocolitis

Necrotizing enterocolitis (NEC) is one of the leading causes of death of gastrointestinal origin in the preterm infant. It is a devastating disease, and at present, it can be considered as part of a spectrum of diseases acquired in the neonatal period characterized by necrosis of the ileum and/or colon. The incidence is 1–5 per 1000 newborns [24]. In multicenter studies, it has been



Figure 4. Simple abdomino-thoracic radiological study in horizontal position showing a great dilatation of the small intestine. Note the pelvic opacity.

estimated that NEC is present in 7–13% of neonates younger than 33 weeks of gestation and weighing less than 1000 g. Mortality is almost 50%, mainly in those requiring surgical treatment. The majority of cases occur in premature babies and a lower percentage in term births [24].

The intestinal epithelium of premature infants is predisposed to have an exaggerated inflammatory response to bacterial colonization, allowing destruction of the mucosa and damage to mesenteric perfusion. This inadequate inflammatory response triggers the emergence of TLR4 (toll-like receptor 4), a response receptor found in the premature gut epithelium that recognizes the lipopolysaccharides of membranes of Gram-negative bacteria. This regulation of the signaling pathway includes nuclear factor kappa beta 1 and IL-1 receptors. Other important risk factors that have been implicated in its development include intestinal immaturity, inadequate bacterial colonization of the intestine, asphyxia, anemia, presence of congenital defects such as gastroschisis and persistent ductus arteriosus, use of medications such as indomethacin, low birth weight, Apgar score of five or less, use of mechanical ventilation, feeding with milk formula and, more recently, the presence of an exaggerated inflammatory response [24, 25].

3.4.1. Diagnosis

The typical newborn with NEC is a premature infant who suddenly develops food intolerance, bloating, bloody stools and signs of sepsis (changes in heart rate, breathing, temperature and blood pressure). An important consideration in the diagnosis of NEC is gestational age and age of presentation. In other words, preterm infants born at 27 weeks of gestation are at greater risk, and symptoms occur at 4–5 weeks of life, compared to term infants. This may be because preterm infants have later colonization of the digestive tract, a prolonged hospital stay and have had broad-spectrum antibiotics used [25].

Bell's classification is widely used to classify the diseases: stage I is non-specific, and stage II is characterized by abdominal distension, wall edema, thrombocytopenia and metabolic acidosis. Radiologically intestinal and hepatic pneumatosis is usually observed. In stage III, signs and symptoms of stage II plus hypotension are present, signs of peritonitis, metabolic acidemia and shock are present. Radiologically, there is pneumoperitoneum (free air in the cavity) as an unequivocal sign of intestinal perforation (**Figure 5**) [25].

Currently, certain biomarkers have been valued in the diagnosis of NEC, which include C-reactive protein and pro-inflammatory cytokines (IL-6, IL-8, TNF- α). In addition, organ-specific markers can indicate damage to the enterocyte as proteins bound to intestinal fatty acids, hepatic, fecal calprotectin and claudin-3. It has been mentioned that the use of Doppler ultrasound is a useful tool to avoid unnecessary surgical operations, since it can verify perfusion of the intestinal wall and eventually identify necrotic intestinal segments before perforation occurs, a benefit that is not obtained with simple radiological studies [26].

3.4.2. Treatment

The initial treatment of patients with necrotizing enterocolitis includes fasting with gastric drainage, broad spectrum antibiotics, intravenous fluids according to the requirements and



Figure 5. Abdomino-thoracic radiography with total abdominal opacity known as “frosted glass sign,” showing enterocolitis in its extreme stage.

water balances, correction of metabolic alterations and adequate management of acid-base imbalances, and inotropic support will be used when there is clear signs of hypoperfusion. Mortality may range from 15 to 63% according to the revised series. Surgery is necessary in more than 50% of the cases, and the objective is to result in the least mutilation possible. Peritoneal drainage without laparotomy may be sufficient and has been reserved almost exclusively for those infants weighing less than 750 g or in those with increased intra-abdominal pressure and compromised ventilation. A laparotomy will be required in 74% of cases. In a typical scenario, there are two alternatives, to perform a primary anastomosis or an enterostomy. In our experience, a primary anastomosis should be performed, particularly if the patient's condition is good and the extirpation of the diseased bowel was extensive (**Figure 6**).

The prognosis of children affected by NEC is characterized by a morbidity of 20–50%; but in developing countries, this percentage is higher, and recurrence after the first event (4–10%), retarded growth (10%), bowel stenosis, mainly of the colon (12–35%), short bowel syndrome (20–35%), neurodevelopmental disorders (30–50%) and stoma complications (50%) are emphasized. The mortality ranges from 15 to 63% according to the series studied [27].

3.5. Neonatal hirschsprung disease

Hirschsprung disease (HD) or congenital agangliosis is a disease of unknown origin, first described in 1888, but it took 60 years for researchers in the field to find the first surgical measure that was more or less effective. This condition is relatively frequent, with one case for every 5000 births and with greater prevalence in males. This abnormality is characterized by the absence of enteric ganglion cells in the distal portion of the colon and, depending on the level of arrest of the neuronal migration, it has been classified as classic, when the ganglionic absence reaches up to 80% in the rectosigmoids and 17% involving sigmoids, splenic angle and transverse colon. The long segment is all forms that go beyond the blind pouch, and total colonic agangliosis is called total agangliosis coli and that which extends from the pectinate line to the duodenum is very rare [28].

Normally, the enteric ganglion cells, which emerge from the neural crest, must innervate from proximal segments of the digestive tract to the terminal colon, and this phenomenon takes place between 5 and 12 weeks of gestation. These cells are responsible for the mobility, not only of the small intestine but also particularly of the colon, which when absent makes the propulsive movements ineffective and the distal intestine function as a deposit. Regarding the etiology and pathogenesis of the disease, many studies have been done; however, conclusions have not been clear. It has been assumed that it appears as a consequence of defects in differentiation due to environmental changes, but there are also studies suggesting that it occurs due to an ectopic expression of class II antigens [29]. Likewise, the influence that genes could have on the disease has been speculated, and it has been found that when the extension is greater,



Figure 6. A transoperative image of the previous case. The contrast of the necrotic intestinal tissue with the normal appearance of the proximal (arrow).

the hereditary tendency increases. Thus, when agangliosis is total, the family tendency may be up to 50%, and if it is in the entire colon, the tendency descends to 15–21%. In relation to chromosomal phenomena, segregation studies have shown that Hirschsprung's disease is a genetic condition transmitted as autosomal dominant, autosomal recessive and polygenic forms. In one karyotype, an autosomal dominant gene was found to cause the disease in the chromosomes 10q11.2. [30]. At the experimental level, it has been found that agangliosis has been able to be reproduced in chicken embryos by extirpation of a segment of intestine, which stopped the neuronal migration [31]. In relation to genetic interference, it is known that the offspring of a woman suffering from the condition is 360 times more likely to transmit the disease than the normal population. The most important constant is that the greater the extension of the aganglionar segment, the greater the risk of inheriting the condition. The conditions that usually accompany HD are neonatal appendicitis [32] and those it shares embryological origin with, such as Wardenburg syndrome, neuroblastoma, pheochromocytoma and Ondine's disease.

3.5.1. *Diagnosis*

Although HD can occur in non-newborn infants [33], it usually manifests and is frequently identified in the first days of postnatal life. An antecedent of delay in the first meconium emission is useful information, a fact that is usually present in more than 90% of the cases. In the case of a neonate who emits his first evacuation after 48 h of postnatal life and with the passage of days suffers constipation, it must be assumed that the cause is congenital agangliosis. Vomiting and abdominal distention are also typical, causing discomfort that can be improved with the application of a suppository or a probe. After showing improvement, they relapse. They are only able to be well for a few days and then suffer from the same picture. There is usually diarrhea, which is almost unequivocally a manifestation of enterocolitis, the most frequent complication of the condition. Radiological study, according to the majority of the authors, is very useful in more than 80% of the cases. A vertical study is recommended, and several findings can be highly suggestive: hydroaerial levels, edema of the intestinal wall, total pelvic opacity and intestinal dilatation. With these data, the next step is to perform the colonic enema (**Figure 7**), which should be done with water-soluble material, without previous enemas and with the introduction of a Nelaton tube no more than 1 cm deep and manually gently inject the contrast medium, while obtaining an x-ray in the lateral position. Images such as jagged rectal wall, a transition zone between a contracted narrow distal portion and a proximal dilatation are highly suggestive. A late study is very useful at 24–48 h. The golden rule for making the diagnosis is undoubtedly rectal biopsy. Performed during or prior to the operation, it will reveal the absence of ganglion cells. Manometry is often useful, especially in older children. One should be very careful to not transgress the normally aganglionar area (a distance from the pectinate line of 10 mm) when performing the biopsy as it leads to false-positive interpretation.

3.5.2. *Treatment*

Non-surgical management is important prior to any procedure, since some of the parameters of the metabolic state of the neonate must be restored. Once the diagnosis is established, a transrectal catheter should be introduced if the patient is very distended and uncomfortable.



Figure 7. The colon shown by enema in lateral position. The spastic area is indicated by an arrow, which is equivalent to the aganglionic segment.

Antimicrobials are indicated as well as intravenous solutions suitable for the patient's age and condition. De la Torre and Ortega [34, 35] have proposed the surgical method of choice in these children, which should ideally be done during the first week of life and in a single operation, that is, without protective colostomy, in order to avoid the disadvantages of bacterial colonization. Unfortunately, this operation requires several requirements so that it can be implemented and three are the most important are to have a confirmed diagnosis within a few days of postnatal life, not to have an aganglionosis coli and to have a reliable pathologist, as much of the success of the procedure depends on them. It is widely demonstrated that at that age and with those requirements met, success is assured in more than 80% of the cases. The classic operations, Swenson, Duhamel and Soave, are left for later. The complications are diverse and include among the mediate complications, stenosis and fecal incontinence, and among the immediate ones, toxic enterocolitis, which is the one most responsible for death. It appears almost indistinctly as a violent infectious diarrheal syndrome, which immediately aggravates the condition of the baby, requiring a neonatal intensive care unit for correction, since the risk of death is very high.

3.6. Gastroschisis

Gastroschisis is a congenital defect of the abdominal wall consisting of evisceration of the midgut and other intra-abdominal organs, such as the stomach, through a small defect of 2–8 cm usually located to the right of the umbilical cord and in direct contact with the amniotic fluid, which gives them the characteristic serous aspect. It occurs between 5 and 10 weeks of gestation and can be identified in utero from week 20. The pathogenesis is still uncertain, with multiple theories. However, it is mentioned that it can be the result of a hemorrhage or thrombosis, which causes an alteration of the flow during the closing process of the wall, causing the exposure of the intra-abdominal organs. A significant increase in global prevalence has been observed in recent decades and ranges from 3 to 20 per 10,000 live births. This may be due to epigenetic changes [36, 37].

Multiple risk factors are mentioned: such as a mother younger than 20 years old or an adolescent, intake of certain medications such as acetylsalicylic acid, pseudoephedrine or ibuprofen, use of drugs, alcohol and tobacco, use of hormonal contraceptives, anemia, exposure to aromatic hydrocarbons, urinary tract infection and, in recent years, the intake of omega-6 fatty acids and alterations in lipid metabolism and pro-inflammatory cytokine substrates [38, 39].

Often the form of presentation of gastroschisis is sporadic and simple, that is, without major or minor malformations, but in a small percentage of cases, it may be part of other syndromes or chromosomal alterations, as well as other conditions such as skeletal dysplasias, Hirschsprung's disease, hydrocephalus and heart disease [40].

3.6.1. Diagnosis

Once the product is born, by either delivery or cesarean section, a preterm newborn between 35 and 37 weeks of gestation with low weight for gestational age is usually identified. In most cases, basic resuscitation is performed, and with respect to the local management of the defect, it is essential to immediately cover the exposed organs with a sterile, non-adherent, resistant, flexible and soft material to reduce loss of fluids and exposure of the peritoneal serosa to the environment. Avoid covering the exposed organs with gauze or compresses [41]. The most severe form of this malformation is a variety called "closed gastroschisis" (**Figure 8**), through the apparently integral abdominal wall, or with an millimetric orifice to the right of the umbilical cord, there is a black or green structure, turgid or fibrous, pediculate and mobile, of a soft consistency, 2–5 cm in length corresponding to the terminal ileum, cecum, cecal appendix and obliterated right colon. This segment is anchored to a fine fibrovascular network that, like a mesentery, keeps it irrigated. There is also intestinal malrotation and an umbilical membrane that is directed proximally toward a very dilated segment that sometimes ends in jejunal atresia variety IIIA. The other end of the digestive tract corresponds to the left colon, which is also obliterated. This occurs between 4.5 and 9% in cases of gastroschisis. There is always almost total absence of the jejunum and ileum [42]. Another variety is evanescent gut. In these cases, the gastroschisis is identified prenatally and at the moment of birth the abdominal wall is integrated; however, the patient presents intestinal occlusion data that requires an exploratory laparotomy that finds jejunal atresia with no more than 20 cm of small intestine and left colon. These children, in our experience, have a fatal outcome.

3.6.2. Treatment

It is important to initiate large amounts of intravenous fluids, electrolytes and antibiotics (**Figure 9**), place a 10–14 gauge Fr. orogastric probe, perform an evacuation enema with a warm solution at 10 ml/kg, avoid hypothermia, correct the acid-base state and, if necessary, provide ventilatory support. Regarding surgical treatment, there are two options: in the case of non-complex gastroschisis, the first alternative is umbilicoplasty, the introduction of exposed organs into the abdominal cavity without enlarging the defect, suturing the aponeurosis and skin and preserving the umbilical cord, preferably in the first 24 h of extrauterine life. We recommend that it should be attempted in all cases. However, success depends on whether there is viscerο-abdominal disproportion, and therefore, it would not be possible to introduce all the intestinal loops or if there is compartmental syndrome. If so, a gradual, delayed closure is preferred between 6 and 10 days, covering it with a PVC membrane or a prefabricated silo. In a comparative study of the two surgical techniques, we found no difference, except that with umbilicoplasty, in-hospital stay time and that the days that the patients required mechanical ventilation and total parenteral nutrition were shorter than with delayed closure [6]. The complex varieties of gastroschisis always require a laparotomy, removal of the necrosed segment or atresia and a primary anastomosis between the residual and viable segments [42].

The prognosis in non-complex cases is generally good with a survival rate greater than 90%; however, in developing countries, mortality may be up to 60% of cases. The main complications are related to prematurity, sepsis, ischemia and intestinal perforation, renal insufficiency or multiple organ dysfunction as well as difficulties for enteral feeding. The prognosis in closed gastroschisis and evanescent intestine is fatal, and both varieties lead to short bowel syndrome, cholestasis, liver failure, sepsis and death.



Figure 8. The clinical aspect of a “closed gastroschisis” in which should be noted the absence of parietal defect, necrosis of the middle intestine and normal aspect of the distal colon, what we call “Antenatal-Paraumbilical-Intestinal-Strangulation” (APIS).



Figure 9. A clinical picture immediately after birth, in which evisceration of the stomach, a good part of the middle intestine and a small portion of the colon can be observed. There is no great serositis because the defect occurred late in gestation.

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