Gynaecol Perinatol 2014;23(1):6–13

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CONVENTIONAL RADIOLOGICAL DIAGNOSTICS OF THE MOST COMMON PATOLOGICAL CONDITIONS OF THE GASTROINTESTINAL TRACT IN NEWBORNS

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Review

Key words: newborn, patological disorders of gastrointestinal tract, radiological methods

Summary. The gastrointestinal tract in neonates has its own anatomical and functional characteristics that are different from those in older children and adults. The most common patological disorders that present in newborn period include developmental anomalies, conditions resulting from the immaturity and genetic diseases. For the purpose of more accurate and well-timed diagnosis of these disorders, close co-operation between clinician and radiologist is needed. In this article we present an overview of the most common pathological conditions and available conventional radiological methods.

Introduction

Radiological diagnostic of the newborn gastrointestinal system demands good knowledge of anatomical and physiological characteristics, as well as of conventional radiological methods. The choice of adequate radiological methods for individual pathological condition to get the answer for targeted clinical question avoids unnecessary accumulation of examinations, what is enabled by close cooperation between clinician – pediatrician neonatologist and pediatric surgeon with radiologist. Performing radiological examination is justified exclusevely by impossibility to prove pathological condition with another method more conservative for the patient.[1]

Neonatal or newborn age is defined as a period from birth till 28th day of life. Anatomical and physiological characteristics of the newborn gastrointestinal system differ from those of infant and older child. Esophagus is quite moveble, with less pronounced mucosal folds and peristalsis. Stomach is horizontally laid, larger in regard to other parts of gastrointestinal system, often expanded by big amounts of gas. Duodenal bulbus is relatively small, with scarce musocal folds. Intestine goes through largest changes in embrionic period; rotation and fixation. Small intestine have poorly expressed mucosal folds, peristalsis is vivacious, junction between jejunum and ileum as well as ileum and cecum is inconspicious and hard to differentiate. Sigmoid colon and rectum are long and movable, colon has less prominent haustration.

In normal conditions, 10 minutes after birth and first breath there is air is in the stomach, after 30 minutes in duodenum, in ileum in 3 hours and after 5 to 12 hours reaches the rectum.[2] Distension of stomach and small intestine is normal in newborns as a result of lying position and increased swallowing of the air during crying and eating.

Conventional radiological methods used in pathological conditions of the newborn gastrointestinal system are: plain abdominal radiography, contrast gastrointestinal series (esophagus, stomach and small intestine), contrast enema. Because of his unharmfulness and easy performance in neonatal intensive care units ultrasound can give some additional useful informations that contribute to accurate diagnosis.

Plain radiography is performed in lying or hanging (standing) position. Contour of abdomen, soft tissue shadows of intraabdominal organs, gas distribution and distension of intestinal lumen are analyzed on the image. Abnormal results are signs of pneumoperitonum, pneumatosis of intestinal wall, distension of individual segments of gastrointestinal system *(Figure 1)*. Native radiograph, according to Wangensteen-Rice, in hanging position and sideways with head downwards is used in algorithm for proving rectal atresia *(Figure 2)*.

Gastrointestinal series are performed with application of positive peroral contrast, usually disolved solution of barium sulfate in amount of 100–200 ml. In cases of possible aspiration, suspicion of perforation, atresia and fistule, and in postoperative period, water-soluble low molecular contrasts are used. Aim of examination is analysis of position, peristalsis and movability of intestinal segments till the junction of ileum and cecum.

Contrast enema study is performed by modified method of double contrast, application of barium sulfate solution and air through the catether inserted in rectum. The same contrasts are used as in series, with possibility of use of high osmolar contrast as a therapeuthic method in suspicion.of meconium ileus.

Ultrasound is noninvasive, nonionizing and unharmful diagnostic method, but of limited significance in analysis of the newborn gastrointestinal system. It is used for detection of free intraabdominal fluids, assesment of the size of atretic segment of rectum, and unin-

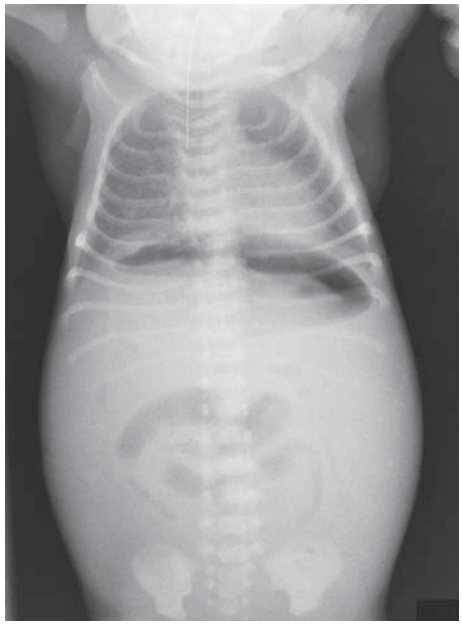

Figure 1. Pneumoperitoneum-collection of free gas under the diaphragm. X-ray babygram

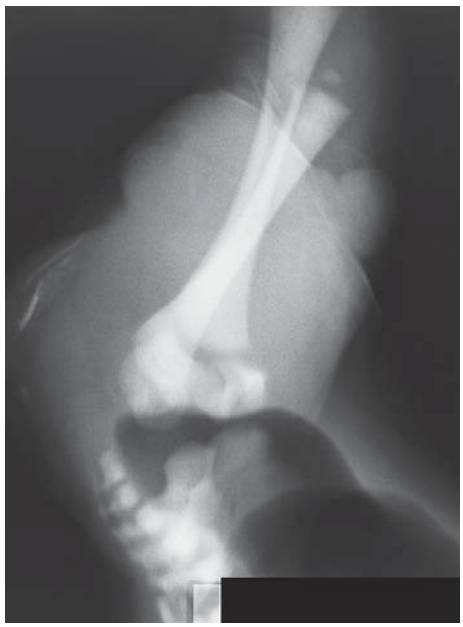

Figure 2. Upside-down lateral x-ray.

voidable method for analysis of hepatobiliary system. In later newborn age it is a method of choice for diagnosis of hypertrophic piloric stenosis, capable to determine muscle thickness and length of piloric canal *(Figure 3)*.[3] Use of doppler gives insight of blood flow in large abdominal blood vessels, and detection of conditions accompanied by vascularization disturbances.

Frequent pathological conditions of gastrointestinal system presenting in the newborn period:

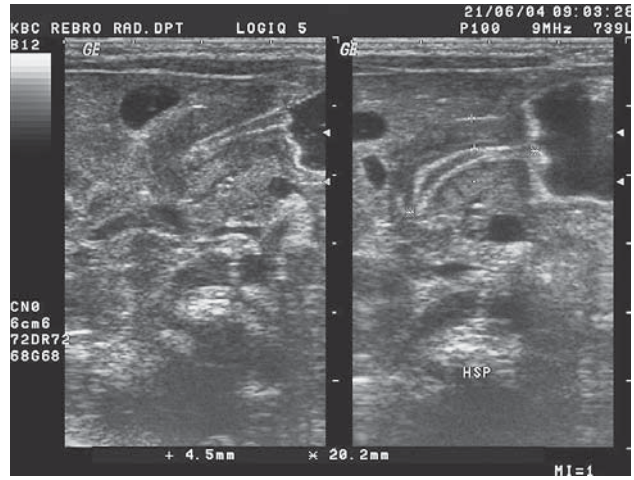

Figure 3. Hypertrophic pyloric stenosis. Ultrasound.

Esophageal atresia and tracheoesophageal fistula

Esophageal atresia and tracheoesophageal fistula are the most frequent congenital opstructive anomalies of the esophagus and relatively frequent malformations in general with incidency 1/3500 live births.[4] There are 5 forms: simple atresia without fistula, atresia with fistula with proximal or distal esophageal pouch, atresia with proximal and distal fistulas, tracheoesophageal fistula without atresia so-called H-fistula.[5] The most frequent position of atresia is upper third of the esophagus. The anomaly occurs more often in children with Down syndrom and as a part of VACTREL syndrom (vertebral

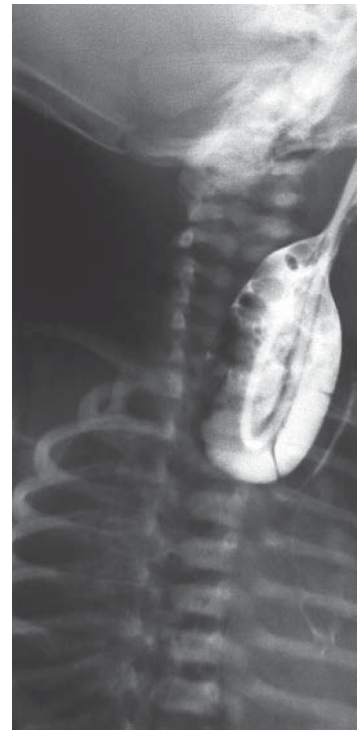

Figure 4. Oesophageal atresia. Nasogastric tube coiled in the oesophageal pouch, isotonic contrast media study.

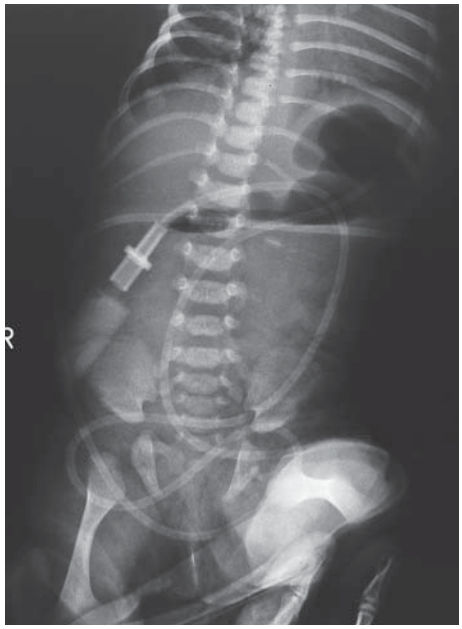

Figure 5. Duodenal atresia. Doubble bubble sign. Plain abdominal x-ray.

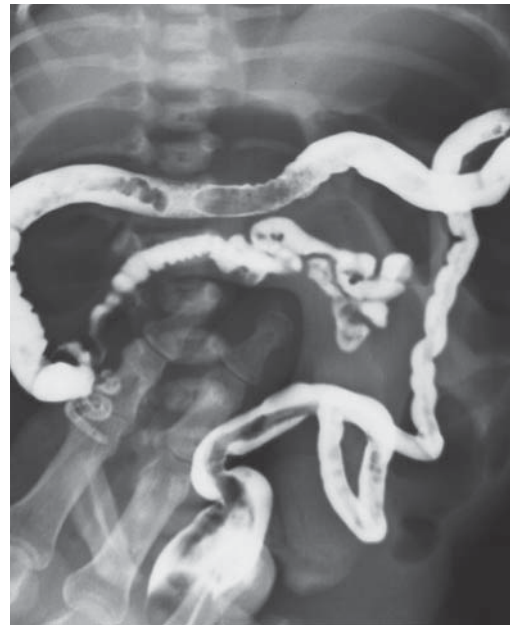

Figure 7. Ileal atresia. Barium enema notice level of ileal atresia with functional microcolon.

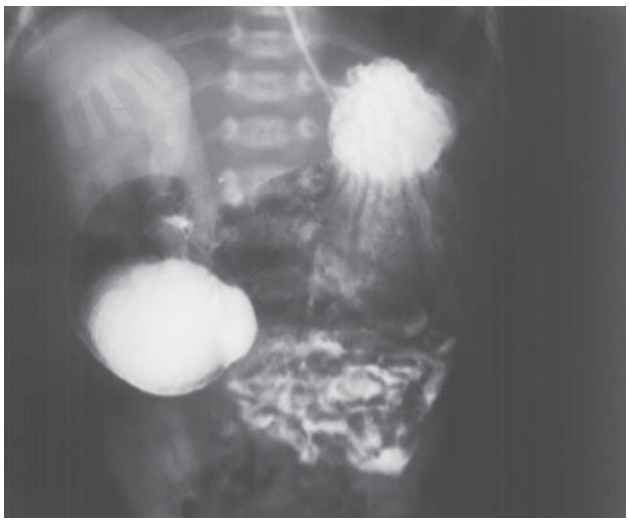

Figure 6. Duodenal membrane with small perforation hole. Barium contrast examination of the upper gastrointestinal tract.

anomalies, duodenal atresia, rectal and renal anomalies..).[6,7] Simple atresia is manifested by enhanced salivation, coughing, choking and cynosis due to aspiration even at first brestfeed. Plain radiograph demonstrates baggy air-filled esophageal pouch, without signs of gas in the stomach and small intestine. In case of atresia with tracheoesophageal fistula, fistula is most often located immediately above tracheal carina. It is manifested as atresia without fistula but on the radiograph stomach and small intestine are filled with gas. Radiographic examination is performed with catheter inserted in esophageal pouch, small amounts of contrast is applicated to define distal border of atresia and contrast is aspirated after the examination *(Figure 4)*. Tracheoesophageal fistula without atresia presents after months or years. Treatment of esophageal atresia and tracheoesophageal fistula is surgical.

Atresia and stenosis of duodenum and small intestine

Duodenal atresia is caused by complete obliteration of intestinal lumen, as a failure of duodenum to recanalize at 6th week of gestation. Duodenal membranes present as atresia. Stenosis is partial obstruction of duodenal lumen caused by either partial failure to recanalize, membrane with perforation or anular pancreas. Atresia and stenosis are usually located in the area of ampulla of Vater; one third above it and two thirds bellow it.[8] Congenital duodenal atresia is 15 times more common than the congenital stenosis or stenosis caused by anular pancreas. Annular pancreas is ring shaped uncinate process caused by embrional development anomaly. It usually causes subtotal stenosis of medial part of descending duodenal segment, and if comlpete it is hard to differentiate it from the congenital atresia. Rarely extraluminal obstruction is caused by preduodenal portal vein.

Incidence of duodenal atresia and stenosis is 1/10 000 live births,[9] equally male and female, more often in children with Down syndrome and as a part of VACTREL.[7,10,11] It is manifested by bilious vomitting in first hours after birth. Bile may be absent in case of preampullar atresia. Stomach and duodenal bulbus are dilated with air-fluid levels presenting characteristic »double-bubble« sign, without gas distally from the place of obstruction *(Figure 5)*. Small amounts of gas may be present distally of the place of obstruction in case of

stenosis. Examination can be supplemented by aplication of positive contrast to exclude distal stenosis. Treatment is surgical.

Total duodenal membrane manifests as atresia and radiography examination is the same, so diagnosis is confirmed intraoperatively. In case of membrane with perforation hole small amounts of contrast are visible in small intestine with gastrointestinal series. Intestine distally of obstruction have narrow lumen as opposed to very distended megaduodenum proximally to membrane (»tenis-racquet« sign) *(Figure 6)*.[2]

Atresia of small intestine is a congenital anomaly presenting as a mechanical ileus. Cause is usually vascular intrauterine incident. It is manifested by vomitting in first hours after birth, like duodenal atresia. Atresia and stenosis of small intestine are most often located in ileum and may be multiple. Contrast defines level of atresia co-presenting microcolon due to out-of-function state. Atresia can be suspected with conventional gastrointestinal series but analysis is hindered by stagnation and dilution of contrast in distended small intestine, so atretic segment is better defined by contrast enema. Treatment is surgical.[6]

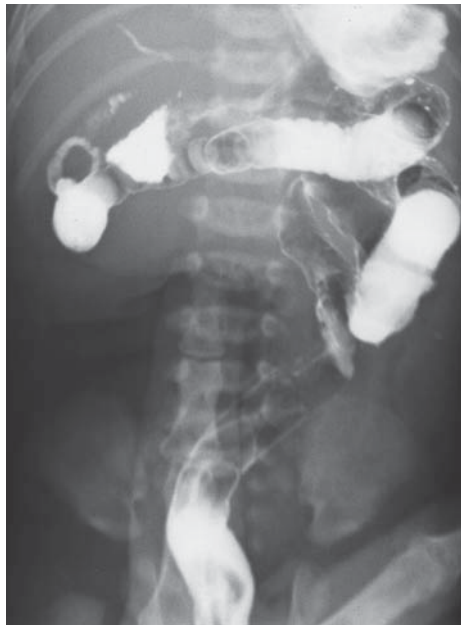

Figure 9. Subhepatic displacement of the caecum. Double contrast enema.

Rotation and fixation disorders

Malrotation is one of the most important cause of duodenal and small intestine obstruction in newborns. Malposition itself does not cause significant simptoms, but joined with malfixation can have drastic consequences. In case of fixation disorders duodenal flexure and cecum are not in their usual location, and the root of mesentery is short. That leads to rotation of intestinal loops around mesentery, that, if prolonged, can further lead to occlusion of vascular vessels and consecutive ischaemia. In some children with malfixation there are additional aberant peritoneal folds so-called Ladd's bands. They connect malpositioned cecum to the lateral wall of right hemiabdomen and compromise horizontal segment of duodenum. These disorders are manifested as simptoms of duodenal stenosis; bilious vomitting, and simptoms of volvulus if it develops. After application of positive contrast compression of duodenum by Ladd's bands is presented as funnel shaped or grooved (»sink« and »groove« signs) *(Figure 8)*. Disorders of rotation and fixation are presented ay conventional contrast radiological methods as atypical location of duodenojejunal flexure right of spine and caudal of usual location. In case of volvulus sign of »corkscrew« is pathognomic; colon si located in left and small intestine in right hemiabdomen, with subhepatic displacement of cecum *(Figure 9)*.[2]

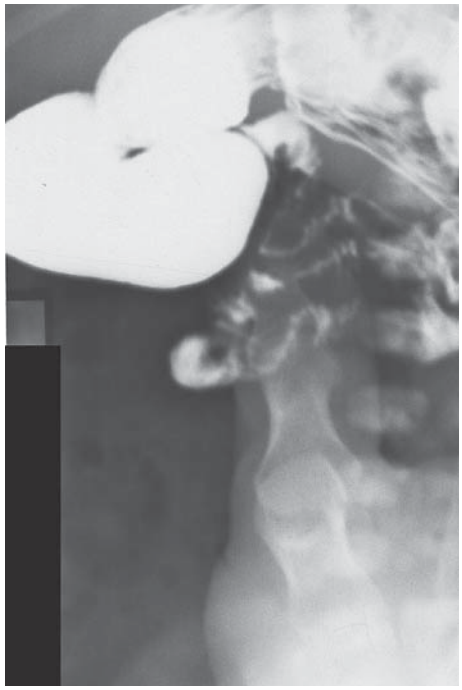

Figure 8. Malrotation of the mid-gut. Peritoneal Ladd's band compression of the duodeunom with narrowing of the gut lumen. Barium contrast examination.

Meconium ileus

Meconium ileus is one of the most common causes of distal obstruction of small intestine as a result of sticky thick meconium stasis in distal ileum. It is usually earliest manifestation of cystic fibrosis,[12] but can be associated with anomalies of pancreatic duct. As a result of unadequate secreation of pancreatic juices meconium is not soft but thick which leads to distal obstruction of small intestine. Consequently lumen of the colon is narrowed – microcolon. Intrauterine complication is intestinal perforation with development of peritonitis and later peritoneal calcifications. It is manifested with vomitting of darkgreen fluids, abdominal distension,

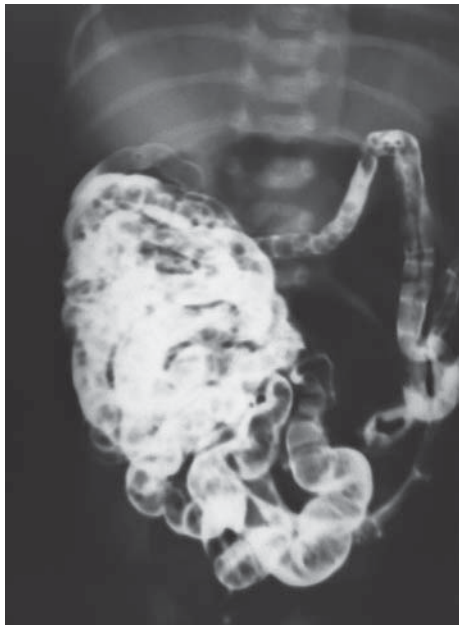

Figure 10. Meconium ileus. Intraluminal filling defects due to meconium. Water-soluble contrast enema with reflux in the ileum.

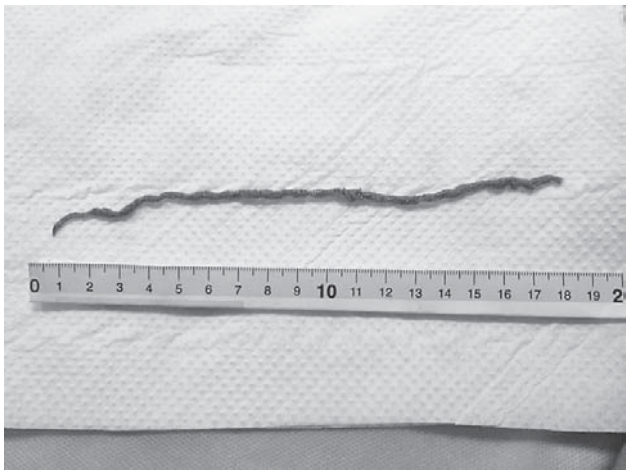

Figure 11. Meconium mass. Therapeutic effect after water-soluble contrast enema.

strong peristalsis and obstipation with unusually small anus and rectum on digitorectal examination. Plain radiograph is nonspecific. There are air-liquid levels and dilatation of intestinal loops, but less expressed than in intestinal atresia. In lower right quadrant »soap bubble« sign may be seen due to mixing of meconium and gas.[11] In children with intrauterine perforation calcifications are spreaded in abdomen. Contrast enema presents microcolon, and reflux of the contrast in the terminal ileum presents intraluminal round-shaped filling defects due to thick meconium *(Figure 10)*. With enema, or gastrointestinal series with water-soluble contrast, either diluted gastrografinum, therapeutic effect might be achieved *(Figure 11)*. In the process it is necessary to achieve reflux to terminal ileum with enema to soften the meconium, while contrast passage has to be performed carefully and slowly in caution of perforation.

Necrotizing enterocolitis

Necrotizing enterocolitis (NEC) is a urgent high-risk idiopathic enterocolitis occuring in prematures and very low birth weight newborns. It occurs as a consequence of hypoxia and hypotension, central redistribution of blood flow and resulting intestinal ishaemia. It begins as a inflammation and edema of intestinal wall that leads to infarction and necrosis with resulting bacteriemia, sepsis and pneumoperitoneum. Despite improved care for premature newborns NEC is still one of the leading causes of their morbidity and mortality. Incidence is 1–5% of newborns in intensive care units and 5–10% of those with very low birth weight. (<1500 grams).[13] Time of occuranse is reversely related to gestational age; premature newborns usually do not develop NEC until second or third week of life, even later.[14] Mortality is 20–40%, increasing with lower birth weight.

Radiological diagnostics with clinical manifestations have important role in diagnosis, follow up of therapy success and diagnosis of possible complications. Oftenly interpretation of plain radiograph is a chalenge., especially in early stage of the disease due to absence of radiological signs. Bell staging criteria classify the disease in three stages: stage I – suspicion of NEC, stage II – diagnosis of NEC, stage III – advanced disease. Distal ileum and proximal colon are oftenly affected, but any segment of intestine as well.

When there is suspicion of NEC plain radiograph is taken every 6, 12 to 24 hours depending on the condition of the child.

The role of the radiologist is to help neonatologist define diagnosis before appearence of specific symptoms.[17] Plain radiograph shows diffuse distension of intestinal loops. Distribution of gas can be nonspecific and assymmetric, with separation of the loops. When NEC is established imaging is more specific. There is accumulation of gas in the intestinal wall in shape of linear, half-moon and oval clusters (intestinal pneumatosis), depending of gas site – subserosal or submucosal *(Figure 12)*. Pneumatosis can occur in newborns without major simptoms, on the other hand newborns with severe or even fatal manifestations may not develop pneumatosis. Except pneumatosis, one of other pathognomic signs of NEC is gas in portal vein. It occurs more often in severe forms of disease and oftenly it is associated with letal outcome.[18,19] On plain radiograph there are transparences branching from portal vein to periphery, and on ultrasound they appear as hyperechogenics moving inside portal branches.

In advanced disease there are necrosis and perforation that manifest as pneumoperitoneum, main indication for surgery. Perforation is indicated by so-called sign of fixated loop; dilated intestinal loop appears rela-

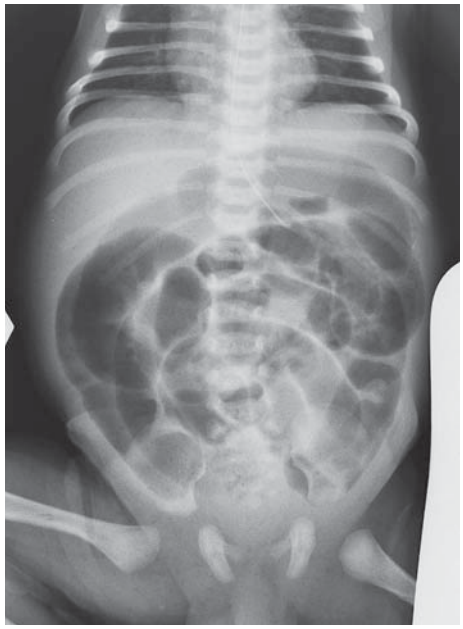

Figure 12. Necrotizing enterocolitis. Gas translucencies in the bowel wall. Plain abdominal x-ray.

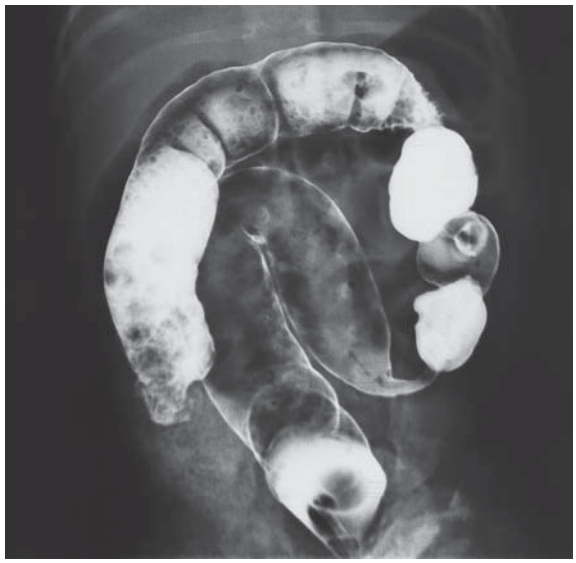

Figure 13. Necrotizing enterocolitis. Colon stenosis late complication. Double contrast enema.

tively unchanged on repeated radiographs in intervals of 24 to 36 hours. In case of perforation there is also change from diffuse to asymmetric distribution of dilated loops, as well as ascites, most easily presented with ultrasound. None of this signs is specific to identify perforation. On plain radiographs pneumoperitoneum is visible in only 50–75% of newborns with perforation. Main challenge and role of the radiologist is to detect perforation and pneumoperitoneum in earliest phase.

Contrast examinations are avoided in acute phase because of possible perforation. Such examinations are indicated in diagnostic processes of later complications; strictures, enterocysts and fistulas *(Figure 13)*.

Hirschprung disease

Hirshprung disease is congenital aganglionic colon resulting fom agenesis of parasympathetic ganglion cells in intramural plexuses. It leads to relaxation disturbances of affected bowel segment and consecutively to obstruction of stool passing, and dilatation of the segment proximal to obstruction. Aganglionar segment always starts in rectosigmoid colon. It can differ in length and affect only rectum and part of the sigmoid (short segment disease), affect segment of colon proximal to sigmoid (long segment disease) or whole colon with segments of small intestine (total aganglionosis). In about third to quarter of cases aganglionic segment is limited to rectum and sigmoid, occuring three times more often in male children.

It is manifested with vomitting and diarrhea, or as delayed passage of meconium and obstipation. Radiological diagnosis is based on contrast enema with diluted barium sulfate, while plain radiograph is nonspecific and without significant importance.

Contrast enema with diluted barium solution shows rectosigmoid in profile projection, with detection of transitional zone and proximal prestenotic segment filled with stool *(Figure 14)*. In normal state, rectum and sigmoid are about the same width and rectosigmoid index is 1, if rectum is narrower and rectosigmoid index is less than 1 Hirshprung disease is suspected.[21] Unregular contractions of aganglionic segment, and abnormal retention of contrast on image taken 24 hours after contrast aplication are visable. In lack of time to develop

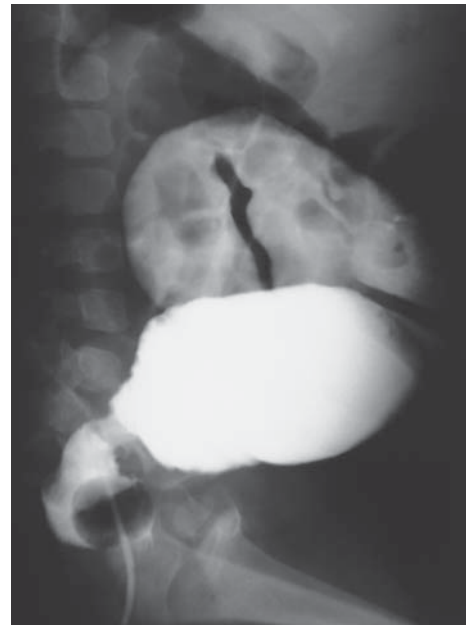

Figure 14. Hirschprung's disease. Aganglionic segment of the rectum, transition zone and proximal dilatataion of the sigmoid colon. Water-soluble contrast enema, profile x-ray.

transitional zone and dilatation of proximal segment in newborns these typical radiological signs are often absent and are noticeable in children of one or two years of age.

Anorectal atresia

There are three types of anorectal atresia: high, intermediary, and low. Exact classification is important because of necessary operative resolution of high atresia. Often fistulas accompany atresia, streching to bladder in male children, and to vagina or bladder in females. Anorectal atresia can be associated with other anomalies as a part of VATER syndrome.

Plain radiograph can be useful only after 5 to 12 hours after birth when air reaches rectum. Imaging according Wangensteen-Rice in profile projection with hanging position with head downwards, is used to detect the level of atresia *(Figure 2)*.[10] Finding of air in the bladder indicate existance of rectovesicular fistula. Fistulas are prooved with fistulography; enema with water-soluble contrast, or during cistouretrography. Ultrasound can determine location of the atresia. Lower level of meconium filled colon is determined and distance to perineum measured. If length is less than 1,5 cm it is low atresia, and if more than 1,5 cm it is high atresia.

Conclusion

Good knowledge of anatomical and physiological peculiarities of the newborn gastrointestinal system is mandatory for timely and accurate diagnosis of patological conditions characteristic for the age. Close cooperation between pediatrician neonatologist and pediatric surgeon with radiologist is important for selection of the optimal radiological method that establishes diagnosis and minimizes the number of examinations.

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Paper received: 14. 02. 2014.; *accepted:* 10. 03. 2014.

KONVENCIONALNA RADIOLOŠKA DIJAGNOSTIKA NAJČEŠĆIH PATOLOŠKIH STANJA PROBAVNOG SUSTAVA KOD NOVOROĐENČETA

Pregled

Ključne riječi: neonatus, patološka stanja probavnog sustava, radiološke metode

Sažetak. Probavni sustav neonatusa posjeduje svoje anatomske i fiziološke karakteristike koje se razlikuju od onih u starijoj dječjoj i odrasloj dobi. Najčešća patološka stanja probavnog sustava koja se manifestiraju u novorođenačkom periodu uključuju razvojne anomalije, stanja nastala kao posljedica nezrelosti i genetskih bolesti. U svrhu što točnije i pravovremene dijagnoze istih potrebna je bliska suradnja kliničara s radiologom. Najčešći poremećaji probavnog sustava u novorođenačkom periodu su atrezija jednjaka i traheoezofagealna fistula, zatim atrezije i stenoze duodenuma i tankog crijeva, poremećaji rotacije i fiksacije, mekonijski ileus, nekrotizirajući enterokolitis, Hirschprungova bolest te anorektalne atrezije. Atrezija jednjaka sa ili bez traheoezofagealne fistule najčešća je opstrukcijska anomalija jednjaka. Klinički se manifestira pojačanom salivacijom, kašljem, gušenjem i cijanozom već pri prvom podoju, a dijagnozu potvrđuje nativna snimka abdomena s prikazom plina u atretičnom, proširenom bataljku jednjaka bez vidljivog plina aboralnije, dok se isti u slučaju postojanja fistule prikaže u želucu i tankom crijevu. Duodenalna atrezija manifestira se povraćanjem već u prvim satima života koje je najčešće praćeno primjesama žuči, a istu kliničku sliku mogu uzrokovati i membrane duodenuma te rijeđe anularni pankreas i preduodenalna portalna vena. Na nativnoj snimci abdomena vidi se karakteristični znak »dvostrukog mjehura«, nastalog kao posljedica distendiranog želuca i bulbusa duodenuma bez prikaza plina aboralno od mjesta opstrukcije. Atrezije tankog crijeva su kongenitalne anomalije sa kliničkom i radiološkom slikom mehaničkog ileusa. Malrotacija i malfiksacija jedan su od najvažnijih uzroka opstrukcije duodenuma i tankog crijeva kod novorođenčadi. Klinički se očituju povraćanjem s primjesama žuči sve do razvoja volvulusa, a kontrastnim radiološkim metodama prikazuju se atipičnim položajem duodenojejunalne fleksure desno od kralježnice i kaudalnije od uobičajenog položaja. Mekonijski ileus nastaje zbog opstrukcije distalnog ileuma ljepljivim i žilavim mekonijem koji je posljedica neadekvatne pankreatične sekrecije. Radiološki se očituje netipičnom slikom aerolikvidnih nivoa uz distenziju crijevnih vijuga te znakom »mjehura sapunice« u donjem desnom kvadrantu. Nekrotizirajući enterokolitis, urgentni i visoko rizični enterokolitis kod prematurusa i djece niske porođajne mase, vodeći je uzrok morbiditeta i mortaliteta među navedenom populacijom, a zbog nepostojanja specifičnih radioloških znakova pravovremeno postavljanje dijagnoze predstavlja izazov, pogotovo u ranoj fazi bolesti. Nakon postavljenja dijagnoze NEC-a, na nativnom snimkama abdomena vide se patognomonični znakovi: pneumatoza crijeva, plin u portalnom sustavu te znak »fiksirane vijuge« i pneumoperitoneum kao posljedica perforacije crijeva. Hirschprungova bolest označava aganglionozu crijeva koja uvijek započinje u rektosigmoidnom kolonu, a zahvaćeni segment može biti različite dužine. Zbog nemogućnosti relaksacije javlja se slika opstrukcije s posljedičnom dilatacijom segmenata crijeva oralno od mjesta opstrukcije. Metoda izbora je irigografija diluiranim barijevim sulfatom u lateralnoj projekciji, uz detekciju razine prijelazne zone i proksimalnog dilatiranog prestenotičkog segmenta crijeva, koji, zbog nedostatka vremena da se razviju, u neonatusa često izostaju. Za dijagnozu anorektalnih atrezija koristi se nativna snimka u laterolateralnoj projekciji s glavom okrenutom prema dolje na kojoj se odredi razina atrezije. Neki od ovih poremećaja kao atrezija jednjaka, atrezije i stenoze duodenuma te anorektalne anomalije češće se javljaju u sklopu VACTREL i Down sindroma.