Complications were recorded until the 30 th postoperative day.

Minor complications, defined as any deviations from normal postoperative course with or without administration of pharmacological treatment, are the most common (infections, metabolic, hematological disturbances).

About a quarter of patients had severe complications, defined with uni or multiorgan dysfunction, need for reoperation and death, which is still high.

Predictor factors for severe postoperative complications were very low birth weight (BW<1500 g), GA < 32 weeks and abdominal operations.

This single centre retrospective analyses show that very low gestation age, very low birth weight and abdominal surgery present risk factors for severe postoperative complications in our NICU patients.

120 TIME TO FIRST PASSAGE OF MECONIUM IN 316 IRISH-BORN TERM INFANTS

Aisling Byrne*, D Cinelli, G Avalos, C Chan, H Cotgreave, S O'Brien, E Moylett, N Abidin.
Galway University Hospital

10.1136/archdischild-2021-europaediatrics.120

Study Background Delayed or absent passage of meconium can suggest an underlying medical condition in a neonate, including Hirschprung Disease, metabolic and congenital abnormalities. Kramer et. Al in 1995 found that 94% of infants passed first meconium within 24 hours. More recently, Ezomike et. al in 2019 demonstrated that 56% of Nigerian infants passed their first meconium within 24 hours. With a discrepancy in the current literature, and recent changes in lifestyle and perinatal factors, it is of interest to detect the average time of first meconium in infants in the developed world, and to assess if this has changed.

Objective To develop a validated reference for average time to passage of meconium in healthy term infants in the developed world, with a higher rate of c-section ($\geq 30\%$) and breastfeeding rate of 60% on discharge. A prospective assessment of time to first meconium passage including the maternal, perinatal and neonatal factors which may influence it.

This prospective cross-sectional study was performed in a tertiary referral center in Ireland. Mothers of term infants in the post-natal ward were interviewed, regarding the timing of first meconium passage, and time of first feed. Any remaining data relating to maternal and infant factors were collected from the medical chart. A questionnaire form was completed by trained data collectors.

Preliminary results include 102 neonates with a mean gestation age of 40 weeks. There is a 55% rate of vaginal delivery and 45% rate of C-section.

The average time to passage of first meconium is 9.3 hours since delivery, with the range between 0 and 83 hours. 93% pass meconium within the first 24 hours. Mothers typically first feed infants within the first half an hour post-delivery, with the first feed consisting of 63% breastfeeding, 33% formula feeding and 4% mixed feeding. Data collection and analysis is still underway.

Preliminary results indicate a vast majority of neonates in the developed world pass meconium in the first 24 hours. Analysis of the maternal and perinatal factors influencing this is underway.

121 BENIGN SERIAL RIB FRACTURE IN A TERM NEONATE

Vedrana Zubčić Zrnč*, Snježana Gverić Ahmetašević, Jasna Tumbri, Rebeka Ribičić, Lorita Mihovilović-Prajz, Iva Kuliš. KBC Sestre milosrdnice

10.1136/archdischild-2021-europaediatrics.121

Aim The aim of the following paper is to present the case of term neonate with serial rib fractures as an isolated finding after birth.

Methods In the medical literature there have been reported some sporadic cases indicating rib fractures in neonates. All of those include fracture of clavícula in complicated labour or other bone affection in bone mineralisation disorder.

Main Finding Most common factors that can lead to rib fractures are birth trauma, cardiopulmonary resuscitation, and metabolic bone disorder which is mostly seen as osteogenesis imperfecta and osteodystrophy of premature babies.

Presented patient had a brief tactile stimulation with no need for excessive reanimation or even mask ventilation. There was no sign of other bone fracture, other systems were not involved but the newborn had bluish discoloration of whites of the eyes.

Conclusion Isolated form of rib fracture without any other affected bone, including intact clavículas, should be considered as benign sign of birth trauma when there is no pneumothorax involved. Even bluish sclera is not predictive sign of a potential syndrome if there are no other multiple fracture of long bone or malformed bones presented at the time of birth (due to intrauterine old fracture)

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Isolated form of rib fracture without any other affected bone, including intact clavículas, should be considered as benign sign of birth trauma when there is no pneumothorax involved. Even bluish sclera is not predictive sign of a potential syndrome if there are no other multiple fracture of long bone or malformed bones presented at the time of birth (due to intrauterine old fracture)

122 MULTIPLE INTESTINAL ATRESIA WITH COMBINED IMMUNODEFICIENCY DUE TO TTC7A GENE MUTATION – CASE REPORT

Iva Vukšić*, Katja Dumić Kubat, Tomislav Čaleta, Toni Matić, Nevenka Cigrovski, Jadranka Kelečić, Miram Pasini, Tomislav Luetić, Jurica Vuković, Nada Sindičić Dessardo, Ruža Grizelj. Department of Pediatrics, University Hospital Centre Zagreb, University of Zagreb, School of Medicine

10.1136/archdischild-2021-europaediatrics.122

Multiple intestinal atresia (MIA) is a congenital malformation disorder in which atresia occurs at multiple levels throughout the gastrointestinal tract, most commonly small bowel. Frequently it presents as an isolated anomaly, but also as a part of a complex inherited disorder caused by homozygous or compound heterozygous mutation in tetratricopeptide repeat domain 7A (TTC7A) gene. These patients usually have associated mild or severe combined immunodeficiency.

Herein we report an infant with MIA and associated combined immunodeficiency (CID). The female child was born at term by vaginal delivery to a 26-year-old G5P3 mother. The parents are healthy, non-consanguineous couple of Albanian origin. Pregnancy was complicated by polyhydramnios. Fetal ultrasound at 19 weeks of gestation revealed dilated bowel loops. Postpartal abdominal X-ray showed signs of gastric atresia.

Surgery revealed widespread atresias, extending from stomach to cecum, and atrophic microcolon. Multiple small bowel resections with end-to-end anastomosis, and ileostomy were done. Histopathological examination showed pseudostratification of the epithelium, increased enterocyte apoptosis, inflammatory eosinophilic infiltrate in lamina propria, decreased small bowel villi, and disrupted apical-basal polarity. At two weeks of age GI obstruction recurred, and she underwent jejunal resection with end ileostomy. The residual small bowel length measured 45cm. Immunological investigation revealed CID: T cell lymphopenia affecting all subsets with lower B cell, natural killer cell count and impaired mitogen response. She experienced several septic episodes. At two months, allogeneic bone marrow transplantation was successfully performed. MIA-CID was suspected and genetic testing confirmed homozygous pathogenic variant in the TTC7A gene (c.315_318del/c.315_318del) causing premature translational stop signal that result with absent protein production. She is now 4 months old, has failure to thrive and parenteral nutrition-induced cholestasis and continues to be dependent on parenteral nutrition for a short gut.

MIA-CID is a rare hereditary disease with about 50 reported patients in literature. Abundant expression of TTC7A gene in thymus and colon and its critical role in intestinal and immune homeostasis can explain this severe phenotype and almost invariably poor prognosis. Clinical course may be more favourable if the bone marrow transplantation is done in the first three months of life. Therefore, immunological and genetic testing should be performed in every neonate born with MIA.

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KAPOSIFORM HEMANGIOENDOTHELIOMA WITH KASABACH-MERRITT PHENOMENON IN A NEONATE: SUCCESSFUL TREATMENT WITH SIROLIMUS

Iva Vukšić*, Tomislav Čaleta, Petra Džepina, Dorotea Ninković, Andrea Dasović Buljević, Boris Filipović-Grčić, Vesna Benjak, David Ozretić, Ana Petrović Gluščić, Ernest Bilić, Marko Radoš, Nada Sindičić Dessardo, Ruža Grizelj. *University Hospital Zagreb, University of Zagreb, School of Medicine*

10.1136/archdischild-2021-europaediatrics.123

Kaposiform hemangioendothelioma (KHE) is extremely rare, life-threatening vascular tumor with estimated incidence of 0.071 cases per

100.000 children. It is notably associated with Kasabach-Merritt phenomenon (KMP), a condition characterised by

profound thrombocytopenia, hypofibrinogenemia, and elevated markers of coagulation activation (D-dimers or fibrin degradation products). Mortality is highly associated with degree of coagulopathy. Patients diagnosed prenatally appear to have increased disease severity. Optimal therapy for KHE is not known. Oral steroids and vincristine for patients with inoperable tumors is most commonly reported, but is associated with limited response and significant side effects.

Sirolimus (rapamycin), a mammalian target of rapamycin (mTOR) inhibitor, is currently being tested in a prospective phase II clinical trial.

Here we present a full-term newborn prenatally diagnosed with giant vascular tumor, affecting the lateral neck. He was delivered by elective C- section and initially presented with a firm, violaceous neck tumor 14x10 cm.

Following the birth, he rapidly evolved to severe KMP. Despite endovascular embolisation performed immediately after birth, his coagulopathy worsened to life threatening hemorrhage (platelets 8,000/μL, fibrinogen 0.8 g/L, D-dimer >10 mg/L, PT-INR 1.36, aPTT 26 s, Hb 80 g/L, Hct 23%), necessitating aggressive blood products replacement to maintain hemostasis.

MRI and laboratory investigations strongly suggested the diagnosis of KHE/KMP. Biopsy was not attempted because of the potential risk of hemorrhage. He was vitally endangered with rapidly enlarging tumor size compromising airway patency, as well as the worsening coagulopathy triggered by platelet entrapment. As there were no response to corticosteroid and propranolol therapy, we elected to start sirolimus (0.8 mg/m² per dose twice daily) along with tapering parenteral corticosteroids. This therapeutic approach led to remarkable resolution of consumptive coagulopathy with stepwise regression of tumor size. After 14 months of follow-up he remains on sirolimus without toxicity, the tumor is barely visible, and MRI showed remarkable reduction in the tumor size.

Currently, no standard treatment guidelines exist for KHE/KMP because of their rare nature and lack of prospective trials. Our experience adds to a growing body of evidence suggesting that sirolimus in the treatment of high-risk KHE/KMP patients might be an efficacious and safe treatment option.

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VEIN OF GALEN ANEURYSMAL MALFORMATION: CASE SERIES OF 14 NEONATES IN CROATIAN NATIONAL REFERRAL CENTRE FOR NEONATE AND PEDIATRIC INTENSIVE CARE

¹Sindičić Dessardo Nada, ¹Čičak Ana Marija*, ^{1,3}Grizelj Ruža, ¹Vukšić Iva, ¹Benjak Vesna, ¹Dasović Buljević Andrea, ^{1,3}Ninković Dorotea, ¹Čaleta Tomislav, ^{1,3}Filipović-Grčić Boris, ²Perkov Dražen, ^{2,3}Radoš Marko. ¹Department of Pediatrics, University Hospital Centre Zagreb, Zagreb, Croatia; ²Department of Diagnostic and Interventional Radiology, University Hospital Centre Zagreb, Zagreb, Croatia; ³University of Zagreb, School of Medicine

10.1136/archdischild-2021-europaediatrics.124

Vein of Galen aneurysmal malformation (VGAM) is a complex congenital cerebro-vascular malformation. Despite some controversy, the overall prognosis of VGAM has improved over time.

We present a series of VGAM patients with hyper-dynamic heart failure during the neonatal period treated in our Department in the past 20 years.

All cases diagnosed with chorioidal VGAM in a single tertiary centre between 2001 and 2020 were ascertained from