

is used to assess organ dysfunction, including liver. According to this scale, the main criterion for liver damage is an increase in alanine aminotransferase (ALT) activity over 50% of baseline. Also, the enzymatic activity of ALT may be increased in newborns with parenteral nutrition.

Objectives The purpose of the study: to study the clinical and laboratory, instrumental and pathomorphological features of septic hepatitis in newborns.

Methods An analysis of 32 inpatient maps of children (of which 29 - (90.6%) premature in gestation 27 - 34 weeks) with septic hepatitis. The only diagnostic criterion was considered to be a combination of the above components with systemic inflammatory response syndrome (SIRS - systemic inflammatory response syndrome).

Results Hepatomegaly was detected in 25 (78%) patients. Hemorrhagic syndrome was detected in 22 (68.7%) infants, most often in the form of pulmonary or gastrointestinal bleeding. Changes in hemostasis in patients were accompanied by an increase in activated. Splenomegaly was detected in 3 (9.4%) children as a result of hyperplasia of the reticulohistiocytic tissue of the organ in response to sepsis and hepatitis. Conjugated hyperbilirubinemia was detected in all 32 patients (100% of cases). Decreased prothrombin index was observed in 25 children (78%), and increased levels of alanine aminotransferase and aspartate aminotransferase (ALT, AST) in 23 (72%). Hypoproteinemia was observed in 12 children (37.5%). Ultrasound in septic hepatitis in 28 (87.5%) patients was characterized mainly by changes in the liver parenchyma and its vascular system, sometimes in combination with biliary tract lesions in the form of thickening of the gallbladder walls and heterogeneity of its contents.

Conclusions Clinical symptoms of septic hepatitis are accompanied by jaundice, in most children hepatomegaly and hemorrhagic syndrome. The main laboratory criteria for septic hepatitis were: conjugated hyperbilirubinemia (100%), decreased prothrombin index (78%) and increased levels of ALT and AST (72%). The most unfavorable prognosis was observed as a result of penetration of the pathogen into the liver through the umbilical vein. In these cases, morphologically formed necrotic hepatitis with total damage to all parts of the hepatobiliary system. The clinical symptoms of the disease were characterized by severe hepatic insufficiency with impaired basic liver function and very high mortality.

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CHALLENGES IN THE MANAGEMENT OF EXTENSIVE APLASIA CUTIS CONGENITA

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Background Aplasia cutis congenita (ACC) is a rare group of congenital disorders characterised by focal or widespread absence of skin, predominantly affecting the scalp, although other areas of involvement have been reported. The exact pathogenesis is unknown, although several theories have been proposed including neural tube defect, vascular compromise from placental insufficiency, intra-uterine infections, genetic mutations, teratogens, and ischemic or thrombotic events related to fetus papyraceus. ACC patients with large scalp defects, often associated with skull defects and/or exposed

dura and superior sagittal sinus have reportedly suffered morbidity or demised from catastrophic haemorrhage from the sagittal sinus, and/or suffered meningitis. Diagnosis is mostly clinical. There is no consensus for the management of ACC given the rarity and variability in terms of the clinical presentation (site) and extent of ACC. Management approaches include conservative treatment, which involves dressing of the aplastic skin until complete re-epithelialisation, and surgical management, which include excision and closure, skin grafting, local flaps and tissue expansion.

Objectives This report describes the treatment of an extensive case of ACC.

Methods The child was managed by a multidisciplinary team comprising the neonatologist, paediatric dermatologist, plastic surgeon, hand surgeon and geneticists.

Results The areas of involvement included most of the scalp and posterior neck, with sparing of the forehead, anterior chest and abdomen, with small portions of the back. Small areas of ACC at joint creases of lower limbs and bilateral upper limbs and bilateral groin regions were also noted. In all, 33.5% body surface area was involved. Both conservative and surgical approaches were considered. On top of the inherent risks associated with conservative management of ACC, such as wound infection, electrolyte abnormalities, and delayed wound healing, this baby was at risk of potentially catastrophic haemorrhage and meningitis due to the large scalp defect. As such, coverage for the scalp was prioritised. Options for skin cover included allograft versus autograft; latter was preferred due to reduced risk of rejection. This approach was limited by the availability of donor sites due to extensive ACC and risks of donor site morbidity. Cultured epithelial autograft (CEA) was preferred but could only be ready after three weeks of tissue culture. As a bridging measure, artificial dermal matrix (ADM) in the form of Integra (R) was applied to the scalp, and other areas, prior to application of the CEA at about 3 weeks of life. The baby was essentially planned for a 2-stage approach to provide urgent skin cover. The baby also received empiric antibiotic coverage and regular wound dressing with topical antibiotics.

Conclusions Aplasia cutis congenita is a rare and potentially life threatening congenital similar to a burns patient, but can suffer from catastrophic morbidity related to exposed sagittal sinus in cases where scalp involvement is extensive. In such cases, we propose prompt surgical coverage of the scalp. We report treatment of a case of extensive ACC involving most of the scalp, using a 2-stage approach, which first entails the application of Artificial Dermal Matrix (ADM), followed by the application of Culture Epithelial Autograft (CEA) when the graft is ready.

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PHENOMENOLOGY OF PEDIATRIC MOVEMENT DISORDERS: FIRST STEP FOR DIAGNOSIS

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Background Movement disorders are broad group of acute and chronic neurological conditions in children and it has been increasingly recognized worldwide because of disturbance of activities of daily living and quality of life. Because of diagnostic and therapeutic challenges for movement disorders, phenomenology plays the key role in diagnosis of specific