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CLINIC-PHENOTYPIC FEATURES OF TURNER'S SYNDROME IN CHILDREN OF CHERNIVTSI REGION

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Knowledge of genesis and clinical symptoms of disease, which is characterized by low growth and gonadal dysgenesis, is necessary for clear orientation in differential diagnostic aspects of some forms of lymphoma and effective medical and genetic counseling.

Purpose: to asses clinical and phenotypic characteristics Turner syndrome in children of Chernivtsi region.

Material and methods. The study involved nine children with Turner syndrome, aged 3-15 years, who were treated at the endocrinology department of Chernivtsi Regional Pediatric Hospital. We used clinic-genealogical, syndromologic, cytogenetic and statistical methods.

Results. Heredity in families of children with Turner syndrome was not burdened, there were no present cases of stunting in families. Correlation between maternal age and appearance of Turner syndrome in children was not confirmed. The course of pregnancy was complicated in 78% of cases. Phenotype determined following features: short stature within 3-10 percentile of the appropriate age (100%); low hair growth at the nape (89%); broad chest with widely spaced nipples (78%); short neck (44%); deformed ears (44%) and its low location (67%); narrow, convex, with a deep nail bed nails (67%); gothic palate (55%); pteryhium (alary folds on the neck) (33%). On pelvic ultrasound in 78% of cases was detected hypoplastic uterus and appendages. In 44% of cases was diagnosed congenital heart diseases, malformations of the genito-urinary system, defects of the musculo-skeletal system, including kyphoscoliosis (33%); double headed flat (22%); cleft palate and mild (11%), violation of dental row (11%). Changes of CNS included cognitive impairment (44% of cases) and symptoms of nocturnal enuresis (11%). In 6 girls (67%) was diagnosed true monosomy of X chromosome; in 3 (33%) - mosaic form of monosomy (46, XX / 45, X0).

Conclusions. Pathognomonic signs of Turner syndrome are: low growth, broad chest, nipples hipertеloryzm, congenital lymphatic swelling of hands and feet. Children with low growth gonadal dysgenesis, as a group of risk of Turner syndrome, require comprehensive and clinical, genealogic and cytogenetic examination for early diagnosis of disease.

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THE ROLE OF SOME INTRAPARTUM AND POSTPARTUM RISK FACTORS IN THE DEVELOPMENT OF HYPOXIC ISCHEMIC ENCEPHALOPATHY IN TERM NEWBORNS

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Although the risk of hypoxic-ischemic hypoxic-ischemic encephalopathy (HIE) is significantly higher in preterm and low birth weight (LBW) infants, among term infants with an adequate to gestational age body weight this condition is not rare. However, the role of intrapartum and postpartum risk factors for the development of HIE depending on birth body weight are still limited and conflicting.

The aim. To study the role of intrapartum and postpartum risk factors in the development of HIE in term infants with normal and low to gestational age birth weight.

Materials and methods. In the department of neonatal pathology of the Chernovtsy Regional Children Clinical Hospital 41 newborns with HIE have been examined. The first (I) clinical group has been formed from 28 term neonates with corresponding to gestational age body weight. The second (II) group included 13 LBW newborns. The survey results analyzed by nonparametric (Pφ, Fisher's angular transformation) method of statistics.

Results. Newborns from I group were born as a result of pathological delivery (by cesarean section, vacuum-assisted delivery or by the help of drug induction) are twice as likely (32,1% of cases) to the II group (15,4% of cases; Pφ>0,05). Tight nuchal cord with development of the newborns' distress during delivery has been occurred in 25% infants of I group, but only in 7,7% of LBW neonates (Pφ<0,05). Pathological amniotic fluids (thick, meconial or hemorrhagic) were registered in 32,1% cases of the I group versus 7,7% of cases in the II group (Pφ<0,05). Severe asphyxia (1-st minute Apgar score ≤ 4 points) was diagnosed in 22,2% patients of the I group and only in 7,7% of LBW newborns (Pφ>0,05). In the early neonatal period a postasphyctic multiple organ dysfunction syndrome has been developed in 14,3% of cases in the I group and 7,7% of children in the II group (Pφ>0,05), while only representatives of the I group had convulsive syndrome (17,9% of infants) compared with no cases in the II group. According to the ultrasound of the heart, the signs of overload of the right heart chambers have been registered in 28,6% newborns of the I group, but only in 7,7% cases in the II group (Pφ<0,05).

Conclusions. The main risk factors preceding HIE in term neonates with adequate to gestational age body weight are: pathological course of intrapartum period (development of fetal distress due to tight nuchal cord in a quarter of infants, pathological delivery in a third (32,1%) cases of supervision), associated in the every third of newborns with acute severe asphyxia, which often led to the development of cardiogenic complications (28,6%), multiple organ failure syndrome (14,3%) and seizures (17,9%).