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Hemihyperplasia - symptoms and diagnostics on the example of a clinical case

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^{1,2,3}Samarkand State Medical Institute **ABSTRACT:** In 1925 Sympson coined the term "hemihypoplasia", which is completely different from the symptom of hemiotrophy (l). Neurogenic hemihypoplasia is characterized by a symmetrical decrease in one half of the body, face, skeleton and soft tissues, while without disturbing the nervous system (with the exception of rare cases of lesions). Right-handedness or left-handedness, as a functional asymmetry, is normative. At the same time, morphological asymmetry implies a difference in the length and volume of the limb. The clinical symptoms of morphological asymmetries are varied, from uniform side reduction to cross asymmetries. Most scientific literary sources note this congenital anomaly, the reasons are poorly understood (1). The border between the healthy and the affected side runs strictly along the midline, in the form of asymmetry of the skeleton, soft tissues and internal organs. A frequent question for medics is which side is healthy, smaller or larger? In such cases, a slight motor difference comes to the rescue, the worst side is weaker, the jerk reflex is worse, the preference for the grasping reflex, the trampling of shoes on one side. Vegetative asymmetry is usually manifested by thinning of the skin, skin color (lighter or darker). Ripple difference between sides.

KEYWORDS: hemihyperplasia, brain – malformation, morphological asymmetry.

INTRODUCTION

An early study conducted at the Department of Pediatric Neurology by prof. Shamansurov Sh.Sh. (2007) showed the absence of movement disorders on the side of hemihypoplasia, the dissertation work was enlightened, the clinical material was based on the examination of 100 children with

neurogenic hemihypoplasia. The main conclusion of the work done was to prove different types of neurogenic hemihypoplasia, respectively with different neurological symptoms. In the work, dematoglyphics indicators had a specific pattern, and to clarify the type of neurogenic hypoplasia, it was proposed to use electroencephalography data, which allows one to see focal EEG phenomena. Scientists of the European Society of Neurologists are inclined to say that in a normal (healthy) body there is a "smart" program that prevents cell growth, unnecessarily; in children, hemihyperplasia cells on one side of the body cannot stop developing, resulting in asymmetry. Some are inclined to talk about neurogenic hemihyperplasia as a consequence of a malformation of the brain - malformation. A developmental abnormality in the perinatal period can put a high risk of tumor growth on the side of hypertrophy, for example, Wilms' tumor, the appearance of cancer in the kidney (1). The genetic factor is not excluded, the Beckwith-Videmala syndrome is similar in etiology (according to the occurrence in one child per 14 thousand children); the syndrome is caused by an imbalance of genes in the region of chromosome 15, according to the mechanism it is a mutation in the maternal algorithm. Thus, the development of neurogenic hemigiperplasia of the body can be prevented by assisted reproductive technologies. A very interesting and further question for discussion is the indicator of the risk of developing hemihyperplasia in children after IVF (1).

Clinical cases. Child 1y. 4 months., Asatullaeva Anora entered the Department of Pediatric Neurology of the 1st Clinic of the Samarkand Medical Institute for the first time. According to the mother, the child is from the fourth pregnancy (there were twins in the third birth), the previous children are healthy. The marriage of parents is kindred, father and mother are cousins. All three semesters of pregnancy proceeded smoothly, during the period of pregnancy, the mother was registered at the place of residence (in the district polyclinic), but did not receive a full examination, there was no screening study, ultrasound examination of the fetus, there were no specific laboratory tests. The girl was born on time, weighing 4kg 500g, which is a characteristic feature of Beckwith-Videmala syndrome. On the Apgar scale, the child's condition at birth corresponded to 8 points. Attached to the breast (according to the normative charts for managing a healthy child), active sucking of the mother's breast was noted. On the 5th day, the umbilical cord disappeared, the child received preventive vaccines in the maternity ward according to the calendar, without complications. On the part of the medical staff, there were no additional recommendations when discharging from the maternity ward home, this can be assessed as the absence of signs of neurological, somatic disorders during and after birth. The first signs of asymmetry, the parents noticed 3-4 months after the birth of the child, according to the mother, her face began to embarrass her, the cheek on one side was larger, especially when she was feeding the baby. Turning to the doctors, excluding the inflammatory process, it was recommended to consult a dentist, suspicion of improper bite growth, or anomaly in tooth growth. At the same time, the child was growing without any special disturbances, from the side of weight, height, ate well, slept on time. Allergic reactions to food are denied, vaccinations were carried out on time without complications according to the calendar... The child began to hold his head at 2.5 months, was interested in toys, played with joy and recognized the parents, all reflexes were quite active (grasping). By the age of 7 months, the child was crawling on his own, trying to sit. It was during this period that parents, accustomed to face asymmetry, noticed the difference in arms and legs, in terms of volume. The beginning and insignificant asymmetry in the limbs, against the background

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of the general satisfactory condition of the child, was left without the attention of doctors at the place of residence. And only when the girl began to take the first steps, and walk more confidently, the sharp limitation in the right extremities in comparison with the left ones, forced the parents to take the problem more seriously. During the examination, the state of health is satisfactory, the child is active. Breathing even, breathing rate 33 per minute. Heart rate without pathology 125 beats. per minute, the abdomen is soft, the position of the liver is within normal limits. Ultrasound examination of internal organs, without pathologies. Urination is normal. Urine analysis, stool within normal limits, regular. Peripheral lymph nodes are not palpable. *The nervous system*. The eye slits are symmetrical, there are no changes in visual acuity (based on the conclusion of the ophthalmologist); co-agreement, nystagmus, anisocoric, ptosis is absent. The reaction of the pupils to light is preserved, the corneal and conjunctival reflexes are caused. The face is asymmetrical, on the left side of the facial features is larger; the cheek on the left catches the eye, it is enlarged, the impression of bloating is created, the corner of the mouth on this side is lowered. The tongue partly falls out of the oral cavity, asymmetric location, taste sensitivity is preserved. Perform mimicry tests (laughter, cry). The right half of the face is smaller in volume, the face of a child, half of "different ages". Hearing is not impaired, swallowing, phonation, speech articulation are not impaired. The child has single stigmas of dysembryogenesis: dysplastic shape of the ears, on the legs between 1 and 2 toes there is a sandal-like gap on the left leg, an asymmetrical skin pattern on the palms. The motor system: active and passive movements of the limb in full, no cuts or paralysis. The tone is normal, the tendon reflexes are preserved, symmetrical, medium liveliness, no pathological reflexes. The volume of the limbs at symmetrical points in three areas give the difference in sides (Table 1). CTITATES

Indicators (and	Diale	T -64
Indicators (cm)	Right	Left
Growth	78 см	
Weight	11 кг	
Head circumference	46 см	
Chest circumference	51 см	
Shoulder volume		
Lower third of the shoulder	11	13
Middle third of the shoulder	13,5	15
Upper third of the shoulder	14	15,5
Arm length	30	31
Leg length	39	41
Brush circumference	10	11,5
Brush length	10,6	12,3
Shin volume		
Lower third of the lower leg	16	18
Middle third of the lower leg	17	20
Upper third of the lower leg	17	20
Thigh volume		

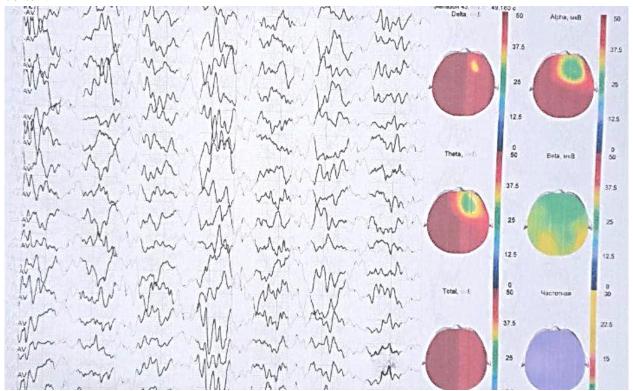
Lower third of the thigh	22,5	26
Middle third of the thigh	23	27
Upper third of the thigh	23,5	27
Foot circumference	10	13
Foot length	12	15,5

Sensitive area (in accordance with possible examination, taking into account age) unchanged. The child walks smoothly, there is no unsteadiness when walking. Stable in the Romberg position. The finger-nose test could not be verified. Visible mucous membranes and skin are of normal color, but there is hyper-pigmentation on the skin in the neck and trunk on the left, on the same side, the skin is dry, the most pronounced dryness on the left leg, the foot on the left looks like peeling like ichthyosis. Psychoverbal development corresponds to age, speaks, understands the addressed speech, from the emotional sphere is noted (according to the mother), frequent mood swings, tearfulness.

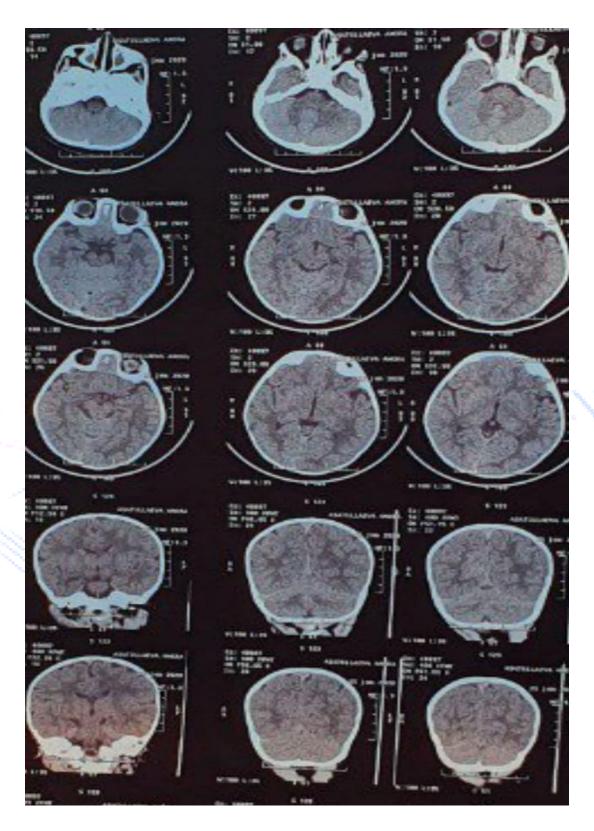
The study of electroneuromyrography gives a variant of the norm.

Doppler ultrasound of peripheral vessels did not reveal pathological or abnormal phenomena.

On electroencephalography, pronounced diffuse changes in the EEG are recorded without signs of local pathology, there is paraoxysmal activity in the form of bilateral synchronous polymorphic waves without a clearly localized focus. Neurophysiological non-maturation of the cerebral cortex.



Computed tomography was taken early before hospitalization does not show pathological changes in the substance of the bra in.



In addition, the child underwent dermatoglyphics, an additional pattern is noted on the tenar of the right palm, it is noted on the third interdigital space of the right hand and has cleft flexion folds of the thumb on the right.



Diagnosis. True congenital right-sided neurogenic hemi-hyperplasia.

As can be seen from the example, the severity of asymmetry increased with age as the pseudo pseudo progression of hemihypoplasia increased. Analysis of anamnestic data (related marriage) indicates the presence of the effect of a mutation during pregnancy; the presence of a developmental anomaly in a child indicates dysembryogenetic etiopathogenesis (malformation has not been proven) of hemihyperplasia; in the recommendations it is proposed to study the genetic spectrum, rehabilitation measures should be according to the scheme in the form of physiotherapy exercises, physiotherapy, and, of course, observation in dynamics.

Conclusion.

The prognosis for children with hemihyperplasia is generally favorable and mainly depends on concomitant abnormalities such as heart failure and swelling. For such children, a neurosonographic and neuroimaging examination of the brain in dynamics is recommended. Early diagnosis of neurological symptoms can prevent the development of the second and third types of neurogenic body hypoplasia in children. The effectiveness of therapy is monitored by anthropolytic research, dermatoglyphic method, and additionally study bioelectric potentials against the background of the EEG phenomenon.

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