https://doi.org/10.54500/2790-1203-2024-3-122-36-41
UDC 61:001.92; 616-091
IRSTI 76.01.39; 76.03.49

Letter to the Editor

Prospects and Relevance of Studying Craniofacial Changes in Anencephaly

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Abstract

Anencephaly, being a rather severe malformation, represents an extensive field for studying such relationships as the brain-skull (brain-base of the skull, brain-vault of the skull, brain-facial skull), brain-endocrine system; not only the theoretical, but also the absolutely complete clinical significance of this kind of research is very great. Of course, first of all, the study of anencephaly covers problems of occurrence (genetic, racial predisposition, diet, metabolic relationships) and diagnosis. However, the cranial changes associated with anencephaly, although primarily described due to the close proximity and relatively easy access, are unfortunately less addressed in the relevant literature. Intensive growth in the technology of plastic and reconstructive operations on the face, orthodontic interventions require thorough knowledge of the formation of skull structures; this formation is impossible without correct relationships with the brain.

The aim of the research was to study the state of research on craniofacial changes in anencephaly. Anencephaly causes significant changes not only in the cranial vault but also in its base and facial part.

A distinctive feature of these changes, first of all, is their extreme variability. The whole variety of developmental processes occurring at the contact between the brain and the skull, with anencephaly, turns into a rather complex malformation picture; studying this picture requires the attention of both clinicians and morphologists. Naturally, all attention is focused on solving etiological and diagnostic issues; in addition, it is necessary to take into account the growing needs of transplantation. But with all this, the anatomical picture of all cases of anencephaly must be clearly clarified, taking into account the cause-and-effect relationships of what is happening, from which it follows that the development of the problem of defects of the primary neural tube in general and anencephaly in particular should be carried out initially from a morphological point of view.

Defects in the central nervous system associated with defects in the formation of the primary neural tube have extremely serious consequences. Anencephaly is the most severe of these developmental defects. Diagnosis at the earliest stages of pregnancy is difficult due to objective circumstances. Even diagnosis within the time limits indicated in the literature, i.e., acceptable for the same objective reasons, requires special preparedness specialists. Considering these associated factors, the study of cranial changes in anencephaly is of particular importance. This concerns both the establishment of certain standards for the development of the skull and the determination of the special properties of the anencephalic skull to improve diagnosis.

Key words: anencephaly, fetus, skull, neural tube defects.

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2024; 3 (122): 36-41
Recieved: 01-08-2024
Accepted: 09-09-2024

Introduction

Absence of the brain (anencephaly) results in acrania (absence of the skull), acalvaria (roofless skull), or cranioschisis (fissured cranium), with variable effects on the face. These fetuses have a minimal survival rate. Ossification of the intramembranous calvarial bones depends on the presence of the brain; in its absence (anencephaly), no bony calvaria forms. In anencephaly, the absence of the calvaria results in cranioschisis, characterized by a short, narrow, lordotic chondrocranium, with notochordal anomalies in many cases; the absence of the brain results in the cranial base remaining unflattened, allowing the facial bones to occupy anomalous positions. Macroglossia typifies anencephaly; trisomy 21 syndrome; Crouzon syndrome; and the association of coloboma of the eye, heart anomaly, choanal atresia, retardation, and genital and ear anomalies (CHARGE syndrome) [1].

Among the neural tube defects, the reported prevalence of anencephaly was highest at 2.1 per 1000 births (95% CI, 1.6–2.8), followed by spina bifida at 1.9 per 1000 births (95% CI, 1.4–2.7). Anencephaly is a neural tube defect that occurs due to the failure of normal tube closure at the cranial end of the 4-week old embryo, resulting in the absence of a major portion of the brain, skull, and scalp. Anencephaly (a Greek word meaning "no brain") is the end stage of a neural tube defect, starting with the (partial) absence of the cranial vault (acrania) (Figure 1). Anencephaly is characterized as the most severe form of neural tube defect (NTD), and the role of folic acid deficiency in its development is discussed. Hispanics are indicated as a population at greater risk in terms of anencephaly development. Also, when discussing other risk factors, attention is paid to defects in folic acid metabolism, obesity and diabetes, dieting, etc. [2–5]. General interest in the topic of anencephaly has increased due to the possible use of anencephaly organs for transplantation and the ethical and scientific issues raised by this possibility [6].

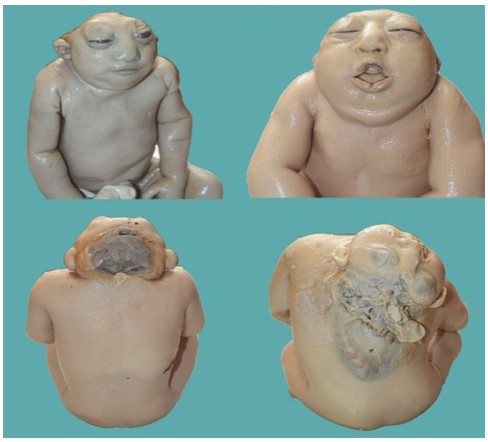

Figure 1 - Anencephaly (from the museum of the Department of Human Anatomy and Medical Terminology of the Azerbaijan Medical University).
*The draft of this manuscript has been approved by the Bioethics Committee of the Azerbaijan Medical University**

The worldwide prevalence of anencephaly is high; therefore, clinicians and specialists need to emphasize the importance of prevention strategies as well as control and treatment strategies [7].

Naturally, the strengthening of the diagnostic component of the problem of anencephaly and neural tube defects in general is associated with ultrasound examinations of the developing fetus. However, until certain stages of pregnancy, due to objective reasons, such a study cannot give clear results [4, 8–10].

Fleurke-Rozema J.H. et al. [4] showed that in a country where first trimester ultrasound at 11 to 14 weeks' gestation is not performed routinely, many cases of anencephaly remain undetected until the midtrimester scan. Anencephaly and exencephaly can be diagnosed in the first trimester, when the skull is not visible due to the absence of the cranial vault, but the face itself, including the orbits, can be visualized. When the brain remnants appear flat, the term anencephaly is used. When brain remnants appear as an irregular, bulging structure, the term exencephaly is usually preferred. Prenatal diagnosis is obvious in the second trimester. In the first trimester, the rounded structure corresponding to the exposed brain can be misleading if an ultrasound is done too early, at 8–10 weeks. This emphasizes the need to perform ultrasounds in the first trimester of pregnancy, at 12–13 weeks, when it becomes possible to analyze the anatomical structures of interest [8].

Exencephaly, anencephaly, meningoencephalocele, and alobar holoprosencephaly were fully detected on scans in the first trimester. Several types of central nervous system malformations may be partially detected on scans in the first trimester, including posterior fossa (PFA) anomalies, open spina bifida, semilobar holoprosencephaly, and severe ventriculomegaly [9]. Anencephaly occurs when the head of the neural tube does not close, resulting in the absence of the fetal skull and brain. By the end of the first - beginning of the second trimester, a normal fetal head should be visible on a prenatal ultrasound. Anencephaly is diagnosed when no calvarial vault or normal brain tissue is visible above the orbits. Careful knowledge of normal intracranial anatomy and the use of a logical sonographic approach can improve the description of abnormalities, leading to a more accurate differential diagnosis in early pregnancy [10].

According to Thomas J.A. et al. [6], for the neuroendocrinologist, the tragedy of the human fetus with the congenital absence of the brain provides at least the opportunity to obtain information about the role of the hypothalamus and its hypophysiotropic hormones in the development of the human anterior pituitary gland and its endocrine target glands. Another issue is the growing evidence that folic acid supplementation around the time of conception reduces the incidence of neural tube defects.

Thus, anencephaly, being a rather severe malformation, represents an extensive field for studying such relationships as the brain-skull (brain-base of the skull, brain-vault of the skull, brain-facial skull), and brain-endocrine system; not only the theoretical, but also the absolutely complete clinical significance of this kind of research is very great. Of course, first of all, the study of anencephaly covers problems of occurrence (genetic, racial predisposition, diet, metabolic relationships) and diagnosis. However, the cranial changes associated with anencephaly, although primarily described due to the close proximity and relatively easy access, are unfortunately less addressed in the relevant literature. Intensive growth in the technology of plastic and reconstructive operations on the face, orthodontic interventions require thorough knowledge of the formation of skull structures; this formation is impossible without correct relationships with the brain.

Based on the above, we set a goal to study the state of research on craniofacial changes in anencephaly.

Craniofacial changes in anencephaly

According to Trenouth M.J. [11], anencephaly has great importance because it acts as a natural experiment for studying normal and abnormal skull growth. Normal craniofacial growth can be explained as a multifactorial process; in this process, all components are balanced and interact in a coordinated manner. This is largely consistent with research [12], which states that comprehensive studies of different bone groups according to their discrete evolutionary precursors, combined with facial analysis, are a vital prerequisite for understanding the interdependence of the development of various tissue components as well as for determining pathogenesis.

Until the studies are compiled, it is difficult to assess which defects are primary and which are secondary.

The specimens in the study of Garol J.D. et al. [13] were classified and grouped as follows: meroacrania, a cranial defect not involving the foramen magnum; holoacrania, a cranial defect involving the foramen magnum; and holoacrania with rachischisis, a cranial defect involving the foramen magnum and extending into the vertebral column. The size of the calvarial defect in fetuses with meroacrania ranged from about one centimeter to several centimeters in diameter, the latter exposing the entire floor of the skull. The opening in the skull was successively limited in front by the frontal bone, laterally by the parietal or squamosal temporal bones, and behind by the parietal or occipital bones, depending on the size of the defect. As the size of the vault defect increased, the size, shape, and spatial orientation of the bones of the calvarium changed more strongly than normal.

Kjaer I. et al. [14] indicated that cases of anencephaly without cervical rachischisis differ markedly from cases with cervical rachischisis. Morphological characteristics, such as bilateral narrowing of the basilar part of the occipital bone combined with normal craniocaudal dimensions, are found in cases without cervical rachischisis. In these cases, frontal clefting of the vertebral bodies was observed. Caudocranial shortening of the basilar part of the occipital bone was found in cervical rachischisis, in which there was also complete median splitting of the vertebral bodies. The study found that when initial closure of the neural groove failed, skeletal abnormalities were more extensive. The study supports the hypothesis that the notochord is an important clue to understanding the pathogenesis of anencephaly.

The cranial floor in cases with meroacrania changed shape from ovoid to trapezoidal, with a narrow end located in the front. This configuration is caused by a decrease in the width of the anterior fossa and the adjacent part of the middle fossa, as well as an increase in the width of the posterior fossa. The middle fossa in front was shallow, and in front of its posterior border, the bottom was not concave but convex. The border between the middle and posterior fossas ran almost perpendicular to the midline. The posterior fossa maintained the same width as the middle fossa instead of tapering posteriorly. In the lateral projection, the bottoms of the anterior and middle fossae were at the same level [15].

Lomholt J.F. et al. [16] noted the importance of the connection between bone compartments and their neural contents. Authors pointed out that neuro-osteologically, the space for the cerebellum is smaller in fetuses with anencephaly than in normal fetuses of the same age, and that there is a difference in size reduction. Examining the development of the cerebellum and brainstem in anencephaly in relation to skull base development may help clarify whether the smaller posterior fossa volume is a developmental error or simply secondary to a calvarial defect. The study revealed two morphological types of the posterior cranial fossa. In one type, the morphology of the cranial fossa was close to normal, whereas in the other type, the posterior cranial fossa was deformed and significantly smaller in size. The latter condition is hypothesized to be due to a primary error in chondral and cranial development.

Because desmal ossification of the neurocranium is induced by the presence of soft tissue (the brain), bone does not develop as a direct consequence of the absence of the brain. The base of the skull, on the contrary, is formed by chondral ossification, which is genetically determined and is therefore also present in anencephaly [17]. The authors indicated that the temporal bone was also one of the bones found in skulls with anencephaly and was positioned vertically in skulls with the foramen magnum and more horizontally in all other skulls without it. From a dorsal point of view, the temporal bones were located at different angles; in skulls without a foramen magnum, their petrous part was located significantly below the horizontal. In addition, the seven skulls had a rather acute angle from a dorsal point of view and less than 150° in the location of the temporal bones, while the two skulls had a rather obtuse angle. Despite the altered arrangement of the temporal bones and the base of the skull, all foramina for nerves and vessels were present, but their position and the direction of the foramina changed. For example, in three skulls, the internal acoustic pore was directed cranially, and in eight skulls, dorsally.

The jugular foramen extended far laterally to the internal acoustic pore. Metzner L. et al. [18] pointed out that in anencephaly, the frontal bone is severely affected. In a normal fetus, the frontal bone forms an angle of 122.3 ± 14.2° with the nasal bone. In the anencephalic skull, there was a marked increase in this angle since the frontal bone does not have an eminence. In anencephalics with meroacrania, the glabellar part of the frontal bone formed an angle of 162 ± 8.7° with the nasal bone and then almost immediately lay flat at an angle of 210 ± 9.8° with the nasal bone. In cases of holoacrania and holoacrania with rachischisis, the angles were 199 ± 4° and 192 ± 7.5°, respectively, for the glabellar part of the frontal bone, and in both groups, the frontal eminence was absent. Morphologically, the most affected facial bone was the zygomatic bone. In the lateral

projection, it usually had a rhomboid shape, but normally it had a "⊥" shape.

The frontal process of the zygomatic bone had a posterosuperior slope, while normally it was directed upward. Consequently, the frontozygomatic suture was located more posteriorly than normal.

As indicated by Trenouth M.J. [11], the nasomaxillary segment in anencephaly was significantly smaller, and the intermaxillary space and mandible were significantly larger than normal. The squamous occipital bone was underdeveloped compared to the norm and had a more vertical slope. During normal growth, the squamous occipital bone rotates from a vertical to a horizontal position as the brain grows. The basilar occipital bone was at a much higher level than normal, and the back of the skull was greatly shortened. The base of the skull was also relatively shorter and at a higher level than the normal standard. This discrepancy was most pronounced posteriorly and decreased anteriorly.

The cranial and facial structures of fetuses with anencephaly were affected in various locations. The most significant changes were observed in measurements related to the transverse plane. All measurements except maxillary length, mandibular body length, and mandibular plane angle differed significantly between anencephaly cases and controls. It turned out that during prenatal development, brain growth prevailed over facial growth. These results indicate that cephalic tissue affected not only the base of the skull but also all facial structures [19]. According to Friedmann I. et al. [20], the anencephalic temporal bones provide an excellent source of comparative anatomy for studying the pathology of Meniere's disease and neurosensory lesions.

The strong association between the cleft palate and the male fetus should be considered during the diagnosis. The presence of associated abnormalities like spina bifida, cleft palate, clubbed foot, clubbed hands, and gastroschisis points to the fact that anenchepaly consists of more than one an etiological entity [21].

As follows from the above studies, anencephaly causes significant changes not only in the cranial vault but also in its base and facial part. A distinctive feature of these changes, first of all, is their extreme variability. The whole variety of developmental processes occurring at the contact between the brain and the skull, with anencephaly, turns into a rather complex malformation picture; studying this picture requires the attention of both clinicians and morphologists. Naturally, all attention is focused on solving etiological and diagnostic issues; in addition, it is necessary to take into account the growing needs of transplantation. But with all this, the anatomical picture of all cases of anencephaly must be clearly clarified, taking into account the cause-and-effect relationships of what is happening, from which it follows that the development of the problem of defects of the primary neural tube in general and anencephaly in particular should be carried out initially from a morphological point of view.

Conclusions

Defects in the central nervous system associated with defects in the formation of the primary neural tube have extremely serious consequences. Anencephaly is the most severe of these developmental defects. Diagnosis at the earliest stages of pregnancy is difficult due to objective circumstances. Even diagnosis within the time limits indicated in the literature, i.e., acceptable for the same objective reasons, requires special preparedness specialists. Considering these associated factors, the study of cranial changes in anencephaly is of particular importance. This concerns both the establishment of certain standards for the development of the skull and the determination of the special properties of the anencephalic skull to improve diagnosis.

Conflict of interest. Not declared.

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Анэнцефалия кезіндегі краниофациалды өзгерістерді зерттеудің өзектілігі мен келешегі

Абдуллаев А.С.

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Түйіндеме

Анэнцефалия дамудың айтарлықтай ауыр кемістігі бола отырып, ми - бас сүйегі (ми-бас сүйегінің негізі, бас сүйегі, ми-бет бас сүйегі), ми - эндокриндік жүйе сияқты қатынастарды зерттеу үшін кең өрісті білдіреді. Мұндай зерттеулердің тек теориялық емес, сонымен қатар толық клиникалық маңызы өте зор. Әрине, анэнцефалияны зерттеу оның себебін (генетикалық, нәсілдік бейімділік, диета, метаболикалық байланыстар) және диагностика мәселелерін қамтиды. Дегенмен, анэнцефалиямен байланысты бас сүйегінің өзгерістері, ең алдымен, жақындық пен салыстырмалы түрде оңай қол жеткізуге байланысты сипатталғанымен, өкінішке орай, тиісті әдебиеттерде аз қамтылған. Бетке пластикалық және қалпына келтіру операциялары технологиясының қарқынды өсуі, ортодонтиялық араласулар бас сүйек құрылымдарының қалыптасуын терең білімді талап етеді. Бұл қалыптасу мимен дұрыс қатынассыз мүмкін емес.

Зерттеудің мақсаты анэнцефалия кезіндегі бас сүйек - бет әлпетіндегі өзгерістерді зерттеудің жағдайын зерттеу болды.

Анэнцефалия бас сүйегінің қоймасында ғана емес, оның негізі мен бет бөлігінде де елеулі өзгерістер тудырады. Аталмыш өзгерістердің айрықша белгісі, ең алдымен, олардың шектен тыс өзгергіштігі. Ми мен бас сүйегінің жанасуында болатын даму процестерінің барлық алуан түрі (ми-бас сүйегінің негізі, бас миы, бас миы-бет сүйегі), анэнцефалиямен өте күрделі даму ақаулық көрінісіне айналады. Бұл суретті зерттеу клиниктердің де, морфологтардың да назарын қажет етеді. Әрине, барлық назар этиологиялық және диагностикалық мәселелерді шешуге бағытталған. Сонымен қатар, трансплантацияның өсіп келе жатқан қажеттіліктерін ескеру қажет. Бірақ мұның бәрімен анэнцефалияның барлық жағдайларының анатомиялық бейнесі болып жатқан оқиғаның себеп-салдарлық байланыстарын ескере отырып, нақты анықталуы керек. Осыдан жалпы бастапқы жүйке түтігінің ақаулары мәселесін, атап айтқанда анэнцефалияны дамытуды морфологиялық тұрғыдан бастапқыда жүргізу керек деген қорытынды шығады.

Біріншілік жүйке түтігінің қалыптасу ақауларымен байланысты орталық жүйке жүйесінің ақаулары өте ауыр зардаптарға әкеледі. Анэнцефалия - даму ақауларының ішіндегі ең ауыры. Жүктіліктің ең ерте кезеңдерінде диагноз қою объективті жағдайларға байланысты қиын. Тіпті әдебиетте көрсетілген мерзімде диагноз қою, объективті себептермен, арнайы дайындықты мамандардың қатысуын талап етеді. Осы байланысты факторларды ескере отырып, анэнцефалиядағы бас сүйек өзгерістерін зерттеу ерекше маңызға ие. Бұл бас сүйегінің дамуының белгілі бір стандарттарын белгілеуге де, диагнозды жақсарту үшін анэнцефалиялық бас сүйегінің ерекше қасиеттерін анықтауға да қатысты.

Түйін сөздер: анэнцефалия, ұрық, бас сүйек, жүйке түтігі ақаулары.

Перспективы и актуальность изучения черепно-лицевых изменений при анэнцефалии

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Резюме

Анэнцефалия, являясь достаточно тяжелым пороком развития, представляет обширное поле для изучения таких взаимоотношений, как мозг-череп (мозг-основание черепа, мозговой свод черепа, мозг-лицевой череп), мозг-эндокринная система; не только теоретическое, но и абсолютно полное клиническое значение такого рода исследований очень велико. Конечно, в первую очередь, изучение анэнцефалии охватывает проблемы причины возникновения (генетическая, расовая предрасположенность, диета, метаболические связи) и диагностики. Однако черепные изменения, связанные с анэнцефалией, хотя и описаны, в первую очередь, из-за непосредственной близости и относительно легкого доступа, к сожалению, менее освещены в соответствующей литературе. Интенсивный рост технологии пластических и реконструктивных операций на лице, ортодонтических вмешательств требуют глубоких знаний формирования структур черепа. Это формирование невозможно без четких взаимоотношений с мозгом.

Целью исследования было изучение состояния исследований черепно-лицевых изменений при анэнцефалии.

Анэнцефалия вызывает значительные изменения не только на своде черепа, но и на его основании и лицевой части. Отличительной чертой этих изменений, в первую очередь, является их чрезвычайная вариабельность. Все многообразие

происходящих процессов развития на соприкосновении мозг - череп (мозг - основание черепа, мозговой свод черепа, мозг - лицевой череп), при анэнцефалии превращается в достаточно сложную мальформационную картину. Изучение этой картины, требует внимания как клиницистов, так и морфологов. Естественно, все внимание сконцентрировано на решении этиологических и диагностических вопросов. Кроме того, надо учитывать растущие потребности и трансплантологии. При всем этом анатомическая картина всех случаев анэнцефалии должна быть четко выяснена, учитывая причинно-следственные связи происходящего, из чего следует, что разработка проблемы дефектов первичной нервной трубки в целом, и анэнцефалии в частности, должна вестись изначально с морфологических позиций.

Пороки центральной нервной системы, связанные с дефектами формирования первичной нервной трубки, имеют чрезвычайно тяжелые последствия. Анэнцефалия представляет собой самую тяжелую из этих пороков развития. Диагностика на самых ранних этапах беременности затруднена ввиду объективных обстоятельств. Даже диагностика в сроки, указанные в литературе, т.е. допустимые по этим же объективным причинам требуют специалистов особой подготовленности. Учитывая данные сопутствующие факторы, изучение черепных изменений при анэнцефалии приобретает особую значимость. Это касается как установления определенных нормативов развития черепа, так и определения особых свойств анэнцефалического черепа для улучшения диагностики.

Ключевые слова: анэнцефалия, плод, череп, дефекты нервной трубки.