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RESEARCH ARTICLE

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# Investigating Craniosynostosis in Infants Resident in Sokoto, Nigeria: A Six Month Prospective Study

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## Abstract

**Background:** Craniosynostosis refers to the premature fusion of one or more of the fibrous joints (sutures) that normally separate the bony plates of infant's skull. This study investigated the prevalence of craniosynostosis in infants resident in Sokoto metropolis.

**Materials and methods:** A total of 51 infants (32 males and 19 females) aged between 0 and 36 months presented at the Institute of Child Health Unit of the Usmanu Danfodiyo University Teaching Hospital (UDUTH) Sokoto were recruited for this study. The head circumference (HC), height and weight of the children were obtained using standard measurements. The 3rd and 97th percentiles of HC were calculated for the study participants using the Standard Infant Head Circumference for Age Percentiles Calculator.

**Results:** Microcephaly was observed in 12.5% of the infants, 1.79% had macrocephaly and 31.25% were normocephalic. The overall mean values for HC was 45.65±3.86 cm in males and 43.37±4.19 cm in females.

**Conclusion:** Craniosynostosis could not be properly established due to non-availability of neuro-imaging records as most of the infants could not afford the cost. Routine neuro-developmental screening including three dimensional CT (computerized tomography) scan and referral of children with Craniosynostosis or abnormal head sizes is recommended.

**Keywords**: Craniosynostosis, head circumference (HC), microcephaly, brain growth, head size, infants.

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## INTRODUCTION

Craniosynostosis is a condition in which one or more of the fibrous sutures in an infant skull prematurely fuses by ossification, thereby changing the growth pattern of the skull [1]. Craniostenosis and craniosynostosis are terms for premature closure of one or more cranial sutures; the result is always an abnormal skull shape [2]. These terms are applicable only to infants in whom the sutures close while the brain is growing [2]. Most cases of craniostenosis are sporadic and of uncertain etiology [2]. Sometimes the resulting growth pattern provides the necessary space for the growing brain, but results in an abnormal head shape and abnormal facial features [1].

Craniostenosis may be one of the features of a larger recognised syndrome of chromosomal or genetic abnormality [2]. Genetic disorders are associated with syndactyly or polydactyly, whereas chromosomal disorders are usually characterised by other limb malformations and growth retardation [2]. In nonsyndromic craniostenosis, the only clinical feature is abnormal head shape [2] . Normal bone growth is impaired in a plane perpendicular to the fused sutures but is able to occur in a parallel plane [2]. In the first years of life the sutures serve as the most important centers of growth in the skull [1]. The growth of the brain and the patency of the sutures depend on each other [3].

Craniosynostosis usually leads to an asymmetric skull (head growth is limited in the plane perpendicular to the fused suture) and to bony ridging overlying the fused suture, characteristics that differentiate it from micrencephaly, where the small head is round, symmetric, smooth, and devoid of bony ridging over the sutures [2, 4].Syndromal craniosynostosis presents with a skull deformity as well as deformities affecting other parts of the body [3]. Clinical examination should in any case include evaluation of the neck, spine, digits and toes [3].

Not all cranial abnormalities seen in children with craniosynostosis are solely a consequence of the premature fusion of a cranial suture [5]. This is especially true in the cases with syndromic craniosynostosis [5]. It is estimated that craniosynostosis affects 1 in 2,000 to 2,500 live births worldwide [1]. Sagittal synostosis is the most common phenotype, representing 40 to 55% of nonsyndromic cases [1]. The second most common type is the coronal synostosis (representing 20 to 25% [1, 6]. The metopic synostosis comes third with 5 to 15% and the lambdoid synostosis is only seen in 0 to 5% of nonsyndromic cases [1]. The common forms of craniosynostosis among syndromes include Acrocephalosyndactyly, Crouzon's disease or craniofacial dysostosis among others [2].

Below are the various classifications and features of craniostenosis [2, 6].

| Terms | Definitions/Descriptions | Mechanism/Cause |
|---|---|---|
| Acrocephaly/Turricephaly | High tower-like head with vertical forehead. | Multiple suture closures |
| Brachycephaly | Broad head with recessed lower forehead. | Premature fusion of both coronal sutures. |
| Oxycephaly (also called acrocephaly in some texts) | Pointed head. | Premature fusion of all sutures (or coronal and saggital sutures). |
| Plagiocephaly | Flattening of one side of the head or occipital flattening. | Premature fusion of coronal or one side lambdoid suture. |
| Scaphocephaly/ Dolicicephaly | Abnormally long, narrow head (parietal protuberance is absent). | Premature fusion of saggital suture. |
| Trigonocephaly | Triangular head with prominent vertical ridge in the mid-forehead. | Premature fusion of metopic suture. |

**Table 1: Table showing classification of cranosynostosis based on shape [2, 6]**

Given the different processes of formation of the various calvarial sutures, it is not surprising that the prevalence of human craniosynostosis detected at or soon after birth is different for each suture [7]. Sagittal synostosis is most common (40–55% of all craniosynostoses), followed by coronal (unicoronal or bicoronal) at 20–25%; the prevalence of metopic synostosis is 5–15% and lambdoid is rare at 0–5% [7]In 5–15% of cases, more than one suture is affected [7].

Head circumference is known to correlate closely with brain volume [8, 9] and can therefore be used to measure brain growth, but a single measurement cannot provide a complete insight into neurological development. Different patterns of early brain growth may result in a similar head size [10]. Results from a study suggest that brain growth during infancy and early childhood is more important than growth during foetal life in determining cognitive function [10].

Microcephaly is a medical condition in which the circumference of the head is smaller than normal because the brain has not developed properly or has stopped growing. Microcephaly can be present at birth or it may develop in the first few years of life. It is most often caused by genetic abnormalities that interfere with the growth of the cerebral cortex during the early months of fetal development [4].

Macrocephaly on the other hand occurs in about 15-35% of autistic children and can also be seen in other types of pervasive developmental disorders, and it is clear that this is the most common physical finding in children with autism [11]. The macrocephaly observed in autism becomes manifest around 1-3 years of age and is typically not present at birth. There is an apparent increased rate of brain growth in the first years of life that diminishes and becomes subnormal in later childhood; macrocephaly in adults with autism is less prevalent than in autistic children [11].

Neuroimaging techniques such as Magnetic resonance spectroscopy has been used to study the biochemical changes that occur in human brain during childhood maturation [12].

In Nigeria, very little work has been conducted with regards head shapes patterns [13] as well as craniosynostosis among Nigerian children in different regions of the country thus the rationale behind this present study.

This study investigated the prevalence of cranioysnostosis in infants seen in the Neurology Clinic, Institute of Child Health, Usmanu Danfodiyo University Teaching Hospital (UDUTH), Sokoto.

## MATERIALS AND METHODS

### Recruitment/Subjects/Population of Study:

A total of 51 infants aged between 0 and 36 months old that attended the Neurology Clinic of the ICH Unit, UDUTH during the period of this study (6 months) were recruited for this study. Verbal informed consent was obtained from parents/guardians of the study participants and the study procedure explained to them before recruitment. The demographic data of the children was obtained from the parents/guardians who brought them to the clinic.

### Inclusion Criteria:

Only infants aged between 0 and 36 months old who visited the Neurology Clinic, ICH Unit, UDUTH, Sokoto during the study period either for routine post-natal check or for any other reason was recruited. Informed consents of parents/guardians were obtained.

### Exclusion Criteria:

All children aged 36 months and above were excluded. Meanwhile, children aged between 0 and 36 months old but with an emergency of life threatening condition were also excluded from the study as well as those children/patients whose parents declined participating in the study.

### Measurement of Head circumference (HC):

The protocol for head circumference measurement used in this study is part of an examination survey on infants and children conducted by The National Health and Nutrition Examination Survey [14]. The circumference of the head was measured on children from birth through 36 months of age under the following steps below to obtain the head circumference measurement.

Position of the study participant: The parent (or guardian) of the infant was instructed to stand holding the child over the parent's left shoulder or else sit with the child in the parent's lap. The parent was asked to remove hair ornaments or braids that might interfere with the measurement. Infants were placed in a sitting position on the parents/guardians lap or, if it is a newborn and has not yet gained head control, supine on the examination table. While older infants > 2 years old were instructed to sit up for the procedure [14].

The Head circumference of the infants < 36 months old was measured using a flexible non-stretchable measuring tape [14]. The head circumference measuring tape was placed around the child's head so that the tape lies: across the frontal bones of the skull; slightly above the eyebrows; perpendicular to the long axis of the face; above the ears; and over the occipital prominence at the back of the head. The tape was moved up and down over the back of the head to locate the maximal circumference [14]. The "zero mark" on the measuring tape was placed on the side of the forehead above the top of the infant's ears and slightly above the supraorbital ridges - the eyebrows. The parents of some of the infants also assisted in holding the tape or comforting the child during the measurement. It was ensured that the tape was firm against the head, and meets the start of the tape on the forehead before measurements were taken to the nearest 0.1 cm [14]. The process was repeated twice for each subject and the average of both measurements recorded on the Proforma.

The 3$^{rd}$ and 97$^{th}$ percentiles of Head Circumference was calculated for the children <36 months children using the Standard CDC/NCHS Infant Head Circumference for Age Percentiles Calculator [15]. Gender-specific z-scores for head circumference were obtained from the Software/Calculator. Each z-score represents the difference between the head circumference of a child and the median head circumference of a reference population (for the same age and sex) divided by the standard deviation of the reference population used by the CDS/NCHS, 2000 [15].

The Head Circumference for Age Percentile Interpretation by CDC/NCHS was used to define the head circumference. Percentile <3 is Microcephaly; percentile >3 and < 97 is normal head circumference; Percentile > 97 is Macrocephaly i.e. Microcephaly as head circumference greater than two standard deviations below the mean and Macrocephaly as head circumference greater than two standard deviations above the mean [15]. The z scores for all the children were calculated.

At the extremes, (>97$^{th}$ percentile or < 3$^{rd}$ percentile), small differences in percentiles represent clinically

important differences in head circumference. At these extremes, the z-score is a more precise reflection of how far the measurement deviates from the mean, and is a useful tool for tracking changes [15].

The postnatal status of the infants was categorized into three as based on previous studies [16]. Infants with normal head size (percentile between = >3 <97) at birth and at follow-up are termed "nomocephalic", infants with small head size (percentile < 3) at birth and at follow-up are termed "microcephalic", and infants with large head size (percentile >97) at birth or follow-up are termed "macrocephalic" [16].

**Data Analysis:**

The data obtained from this study was entered into Microsoft Excel worksheet and analyzed. Statistical analysis was carried out using student t-test to compare the measured parameters between the males and the females. The differences were accepted as significant when $p<0.05$. The analysis was carried out using Statistical Package for Social Sciences (SPSS) version 17 for Windows.

**Ethical Clearance**

Ethical clearance was obtained from relevant Institutions and Departments for this study. Ethical consideration included taking informed consent from parents and guardians of study participants, using Hospital card numbers to maintain confidentiality and making recommendations to appropriate authorities.

**RESULTS AND DISCUSSION**

Out of the total 112 children seen at the Neuropaediatric Clinic of the ICH, Unit of the Usmanu Danfodiyo University Teaching Hospital, Sokoto, 51 (45.5%) were infants aged between 0 and 36 months old. 32 (28.57%) were males and 19 (16.96%) were females.

Moreover, out of the 32 (28.57%) males, 20 (17.86%) were normocephalic, 11 (9.8%) microcephalic and 1 (0.89%) macrocephalic while females had 15 (13.39%) normocephalic, 3 (2.68%) microcephalic and 1 (0.89%) macrocephalic out of the total 16.96% females aged < 36 months (Table 2). Thus, the most dominant head size as per the present study is normocephaliic with a frequency of 31.25% while microcephaly had a frequency of 12.5% and macrocephaly 1.79% (Table 2).

| Head Size Classification. | Males N (%) | Females N (%) | TOTAL N (%) |
|---|---|---|---|
| **Microcephalic** | **11** (9.87) | **3** (2.68) | **14** (12.50) |
| **Normocephalic** | **20** (17.86) | **15** (13.39) | **35** (31.25) |
| **Macrocephalic** | **1** (0.89) | **1** (0.89) | **2** (1.79) |
| **TOTAL** | **32** (28.57) | **19** (16.98) | **51** (45.5) |

**Table 2**: Table Showing the Overall Distribution of Head Sizes in Infants < 36 months old According to Sex.

**Abbreviations: N** = Number; **%** = Percentage

The microcephalic and macrocephalic head sizes observed in this study may be due to the fact that the brain and skull are still rapidly undergoing growth and development into adult size. 90% of which can be attained at 1 year of age [17].

However, microcephaly and macrocephaly seen in age ranges between 1 and 3 years may require further necessary action to determine the etiology. Because it is strongly believed that by age 1 the brain has attained 90% of its adult size [17], thus any deviation from this normalcy might likely point to the need for further investigations including neuro-imagings should be recommended for proper diagnosis as suggested in a study [15]. Craniosynostosis in case of microcephaly and hydrocephalus and autism can be detected using neuro-imagings [11, 12].

| Sex (N) | Max HC (cm) | Min HC (cm) | HC Mean ± SD (cm) | z-Scores | Percentiles | P-value |
|---|---|---|---|---|---|---|
| Males (32) | 50 | 36 | 45.65± 3.86 | 1.669 | 31.81 | **0.0015*** |
| Females (19) | 49 | 38 | 43.37± 4.19 | 1.677 | 33.77 | |

**Table 3:** Table showing the Mean ± Standard Deviation (SD) Values for HC, z-sores and Percentiles for the Mean in Relation to Sex.

**Abbreviations: N** = Number of subjects; **HC** = Head Circumference; **SD** = Standard Deviation; ***** = Statistically significant difference observed between sexes.

The mean HC value in males (45.65±3.86 cm) was higher than that of females (43.37±4.19) and this difference was statistically significant with a p value of 0.0015. The mean HC values in males are greatly higher than the mean head circumference values seen in Turkman males (34.99±1.48) and Fars males (35.10±1.37) [18].

In the present study, the minimum head circumference value in females was 38 cm and maximum value 49 cm, and in males the minimum and maximum head circumference values in this study were 36 cm and 50 cm respectively *(Table 3)*. The maximum head circumference value (50cm) of males in this study is slightly higher than the maximum value (49cm) in females while the minimum value (38cm) in females is slightly higher than that of males (36 cm).

The findings from this study underscores the need for close monitoring of the pattern of growth from birth for the early detection of those with persistent subnormal head size that is either congenital or of post-natal onset. It is pertinent to mention that small head size may also be constitutional and inevitable in some infants and highly correlated with maternal head circumference [19] which though was not elicited in this study. While special efforts were made to minimize measurement errors resulting from possible shape

distortion and scalp edema in the newborns at birth, it was not unlikely that excessive molding in some infants would have contributed to the reported incidence of macrocephaly or microcephaly.

This study has therefore provided valuable information in infants aged between 0 and 36 months old seen in Sokoto, Northwestern Nigeria and this could be used in the diagnosis and treatment in plastic and oral surgery [20] and forensics for the reconstruction of craniofacial remnants [21] and in paediatric neurology.

**LIMITATION OF THIS STUDY**

The sample size used in this study is not sufficient enough to make general conclusions thus, the need for a larger sample size. However, the HC evaluation is a good indicator of skull and brain growth and can be of clinical importance in determining the timing and aetiology of brain insults, especially in the first four years of life.

Routine neurodevelopmental screening including three dimensional CT (computerized tomography) scan and referral of children with Craniosynostosis or abnormal head sizes is recommended

**CONCLUSION**

Craniosynostosis or other causes of microcephalization were not properly established due to non-availability of neuro-imaging records as most of the infants with microcphaly or macrocephaly could not afford the cost of CT-Scans. A statistically significant difference in the mean head circumference was observed between male and female infants.

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