ORIGINAL ARTICLE

Incidence of Undetected Spina Bifida Occulta in Patients Presenting for Other Neurosurgical Conditions in The Emergency Room of a Tertiary Health Facility

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ABSTRACT

Background: Spina bifida occulta are the most common spinal dysraphisms, yet, the least diagnosed. It is estimated that as much as 30–50% of normal healthy people could harbour this defect.
Objectives: To evaluate the incidence of spina bifida occulta and tell-tale signs amongst consecutive patients presenting to a tertiary health facility, under emergency conditions, who had no previously known congenital spinal dysraphisms.
Methodology: The study was conducted at the Accident and Emergency Room of Nnamdi Azikiwe University Teaching Hospital (NAUTH) Nnewi, Anambra State, Nigeria over a 12-month period. The cohort of patients at the Emergency Room were preferred, in order to reduce the interference from overt dysraphisms. Consecutively, consenting patients, were recruited, and data were presented in charts, tables and graphs, and analysed by comparative evaluations with literature.
Results: There were 109 recruited subjects, within the age range of 9 to 85years, and most were males 80 (73.4%). Occult dysraphisms were detected in 8 (7.34%) of cases, 6 (5.5%) of whom were males. The affected spinal regions were: cervical 1 (0.92%), thoracic 1 (0.92%), lumbar 5 (4.6%) and sacral 1 (0.92%); a pre-morbid deficit of unilateral foot drop was found in 2 (1.83%). At presentation, most of the subjects came with neurological deficits; however, their dysraphisms were only discovered at routine imaging. There was no family history of spinal dysraphism amongst them. The tell-tale signs were abnormal hair growth 7 (6.42%), and dermal sinus 1 (0.92%). Spinal magnetic resonance imaging (MRI) and computed tomography (CT) were each employed for 2 (1.83%) cases, respectively.
Conclusion: Occult spinal dysraphisms were discovered in 7.3% of our emergency patients, and most had abnormal bands of hair growth. The non-diagnoses of these cases by relatives and healthcare personnel, prior to their presentation, calls for concern.

Keywords: *Dysraphisms, Folic Acid, Neural Tube Defects, Neuroimaging, X-rays*

INTRODUCTION

Spina bifida occulta result from the incomplete fusion of the neural tube with no overt, or only a very minor defect in the overlying skin. It is reported to be much more prevalent than the open forms of dysraphism.[1,2,3,4,5] Estimates of its frequency come from reviews of spinal radiographs

taken for reasons unrelated to the spine and nervous system.

The incidence of spina bifida with meningocoele or myelomeningocoele is 1-2 per 1000 live births (0.1-0.2%).[6] The risk increases to 2-3% if there was one previous birth, and to 6-8% after two affected children. The risk is, also, increased in families where close relatives like siblings have given birth to children with myelomeningocele, especially if they are from the mother's side. Famine, war and economic disasters show a rise in incidence.[7] The birth-prevalence rate of spina bifida was slightly higher among females than males, with a ratio of 1.2 to 1.[6] In Ile-Ife, Ibadan, Nnewi and Enugu in Nigeria, more males were affected.[8,9,10,11]

Reported prevalence of spina bifida occulta ranges from 5 to 30% worldwide, and it is often, an incidental finding, with an increased incidence of disc herniations.[1,12,13]

Unlike the two other types of spina bifida – cystica and aperta – the occulta is barely detectable unless with experience and a high index of suspicion. And as such, most of the patients with spina bifida occulta may go through life with no neurological deficits.

Its diagnosis is often made by paediatricians on getting a history of subtle but progressive neurological symptoms accompanied by the clinical findings of cutaneous stigmata, such as hypertrichosis, dimples, sinus tracts or capillary haemangiomas, over the spine.[14]

One of the major complications of spinal dysraphisms, especially the occult variant, is the tethered cord syndrome which results from the differential rate of lengthening between the vertebral column and spinal cord, during the growth and development in early life. The vertebral column lengthens faster and farther than the cord.

In tethered cord syndrome, the spinal cord is bound down at the neural placode by the scarred and hypertrophied filum terminale, which would have herniated through the bifid spine defect, making it difficult, if not impossible, for the "bound" or tethered cord to migrate upwards as the vertebral column increases in length. This will, over time, as growth continues, lead to ischaemic and structural damages to the neural bundles of the spinal cord, producing progressive and ascending neurological deficits, in an ascending myelopathy, which are not usually detected early enough.

Inadvertently, it is only when these deficits begin to appear that most patients seek medical help and, those established neurological deficits may be difficult to reverse once they had set in.

Evaluating the incidence of the occult dysraphisms in patients who are presenting with entirely different clinical diseases, like trauma or neoplasms, will, more readily, establish the incidence in our practice and, add to the available body of knowledge.

In Nigeria, and her health institutions, not least, our own tertiary health institution, there is paucity of data on the prevalence or local incidence of this very unique congenital anomaly.

This study would, to an extent, generate our own local data, in this part of the country, by identifying the incidence and major presenting tell-tale signs of spina bifida occulta. Our facility, is a major referral centre for neurosurgical diseases in the South-East and South-South Zones of Nigeria.

The result of this study will provide some baseline data to support the existence of undetected spinal dysraphisms even in this narrow spectrum of patients. It will make a strong case for a wider study on the general population

METHODOLOGY

The study was conducted at the Accident and Emergency Room, Nnamdi Azikiwe University Teaching Hospital (NAUTH) Nnewi, Anambra State, Nigeria. The Emergency Room is a 40-bed section which receives all the emergency cases that present to the 350-bed tertiary facility, and this study was carried out over a 12-month period, 1st November, 2020 – 31st October, 2021. There are both adult and paediatric rooms.

In order to minimize the higher possibility of encountering overt or previously diagnosed spina bifida in the routine outpatient or paediatric clinic, the investigator opted to use the Emergency Room, since the study is focused on emergency care subjects.

It was a prospective, cross-sectional, observational study conducted on consecutive and consenting patients of every age, who were diagnosed with any neurosurgical lesion other than a congenital spinal anomaly.

Data on the presence, or otherwise, of bifid spines in all their spinal imaging studies were collected, then, followed by a direct physical evaluation of the subjects with radiological confirmation of bifid spines, in order to ascertain the presence or absence of tell-tale signs.

Approval was obtained from the Research and Ethics Committee of NAUTH Nnewi.

Collected data were analysed by inferential statistics, and the parameters were compared with the current available data in the existing body of knowledge.

RESULTS

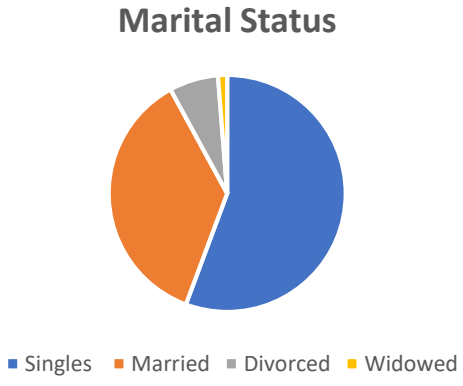
A total of 109 subjects were recruited, with the 0-10year age group numbering 11 (10.1%); whereas >10-20years were 14 (12.8%), >20-30 were 21 (19.3%), >30-40 were 18 (16.5%), >40-50 were 12 (11%); >50-60 were 15 (13.8%); >60-70 were 9 (8.3%); >70-80 were 6 (5.5%); and >80years were 3 (2.75%); *see Table 1*.

Most were males 80 (73.4%); Nigerians 103 (94.5%); and Christians 91 (83.5%). Females were 29 (26.6%), non-Nigerians 6 (5.5%) and Muslims 8 (7.3%), with other religions 10 (9.2%).

Table 1. Age distribution of study population

Age-Range	**Frequency**	**%**
0-10	11	10.1
>10-20	14	12.8
>20-30	21	19.3
>30-40	18	16.5
>40-50	12	11.0
>50-60	15	13.8
>60-70	9	8.3
>70-80	6	5.5
>80-90	3	2.3
>90	0	0

Figure 1. Pie chart showing the marital status of the recruited subjects



The subjects' occupations included the civil service 25 (22.9%), trading 40 (36.7%), schooling 8 (7.3%), menial jobs (Artisans), 13 (11.9%) and dependent 23 (21.1%).
Most of the subjects were unmarried / single 56 (53.4%), while the rest were married 38 (34.9%), divorced 7 (6.42%), and widowed/ widowered 8 (7.33%), *see Figure 1.*

The subjects were those with neurosurgical diseases that excluded overt spinal congenital abnormalities. The presenting lesions were head injury 64 (58.7%), spinal injury 19 (17.4%), hydrocephalus 10 (9.17%), degenerative spine disease 8 (7.34%), intracranial tumour 3 (2.75%), Pott's puffy tumour 3 (2.75%) and subdural empyema 2 (1.83%), *see Table 2.*

Table 2. Distribution of presenting diagnoses

Presenting Diagnosis	Frequency	%
Head Injury	64	58.7
Spinal Injury	19	17.4
Hydrocephalus	10	9.2
Degenerative Spine	8	7.3
Intracranial Tumours	3	2.8
Pott's Puffy Tumour	3	2.8
Subdural Empyema	2	1.8

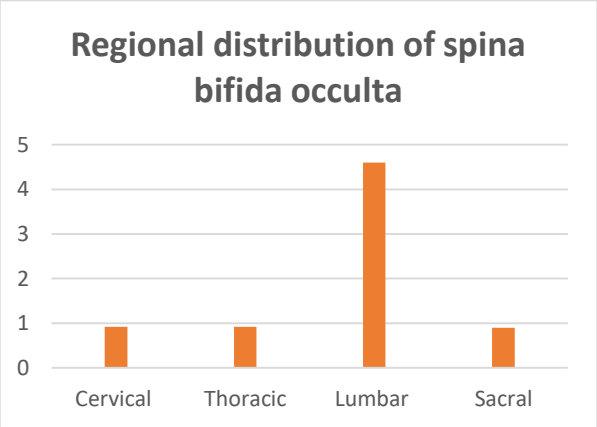
Table 3. Tell-tale signs

Sign	Frequency	%
Band of Hair	7	6.4
Dermal Sinus	1	0.9
Lump	0	0
Mal-pigmentation	0	0
Others	0	0

Amongst the 109 subjects, 8 (7.34%) cases were discovered with bifid spines on imaging, and all were confirmed to have tell-tale signs. Irrespective of their neurological features, it was only those who had bifid spine on imaging that were analysed. These tell-tale signs included: abnormal band of hair 7 (6.42%), and dermal sinus 1 (0.92%); *see Table 3.* None of the 8 subjects had other tell-tale features like lumps or mal-pigmentations.

Six (75%) of the 8 cases were males and 2 were females. The affected regions of the spine were: cervical 1 (0.92%), thoracic 1 (0.92%), lumbar 5 (4.6%) and sacral 1 (0.92%); *see Figures 2-9.*

Figure 2. Bar chart of the regional distribution of spina bifida occulta



Only 2 (1.83%) of the 8 subjects had pre-morbid deficits, which was unilateral foot drop, whereas 6 (5.5%) had no neurological deficits prior to the primary reason for which they had presented to the facility. At presentation, 6 of the 8 subjects came with neurological deficits, whereas the remaining 2 had no deficits; and their lesions were only discovered at routine imaging studies.

There was no confirmed family history of spinal lesions in any of the 8 (7.34%) cases, and, only 2 (1.83%) were aware of their tell-tale signs, while none knew that it could imply an underlying spinal malformation. All the 8 subjects presented with traumatic injuries but, 6 had associated post-traumatic deficits, which included: limb weakness 6 (5.5%) and sphincteric dysfunction in 2 of those 6 subjects. The 6 with limb weakness comprised of 1 each of cervical, thoracic and sacral dysraphisms, respectively, and 3 (of the 5) lumbar dysraphisms. The cervical

dysraphism presented with quadriparesis, whereas the remaining 5 had paraparesis.

An interesting finding was the presence of bifid spines on two different cervical vertebrae (C3 and C7) in one subject, and at two adjacent lumbar vertebrae in another (L4 and L5). Bifid spines were found in the lumbar vertebrae of 5 subjects, 2 of whom had no neurological deficits, despite their dysraphisms and traumatic injuries. Aside the routine spinal x-ray studies, half of the 8 positive cases required further imaging in order to definitively make a diagnosis. Spinal magnetic resonance imaging (MRI), was done on 2 (1.83%), and computed tomography (CT) on the other 2 (1.83%) cases.

DISCUSSIONS

This study identified 8 cases of occult spinal dysraphism amongst 109 cases, which translated to 7.3% incidence rate. This finding falls within the range of 5 - 30% incidence rate reported by various scholars.[1,2,12,13]

Figure 3. Dermal sinus **Figure 4.** Tufts of hair (male)

Figure 5. Cervical 3 & 7 lesions Figure 6. Lumbar 4 and 5

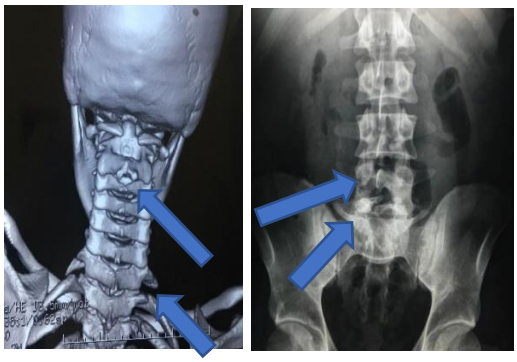
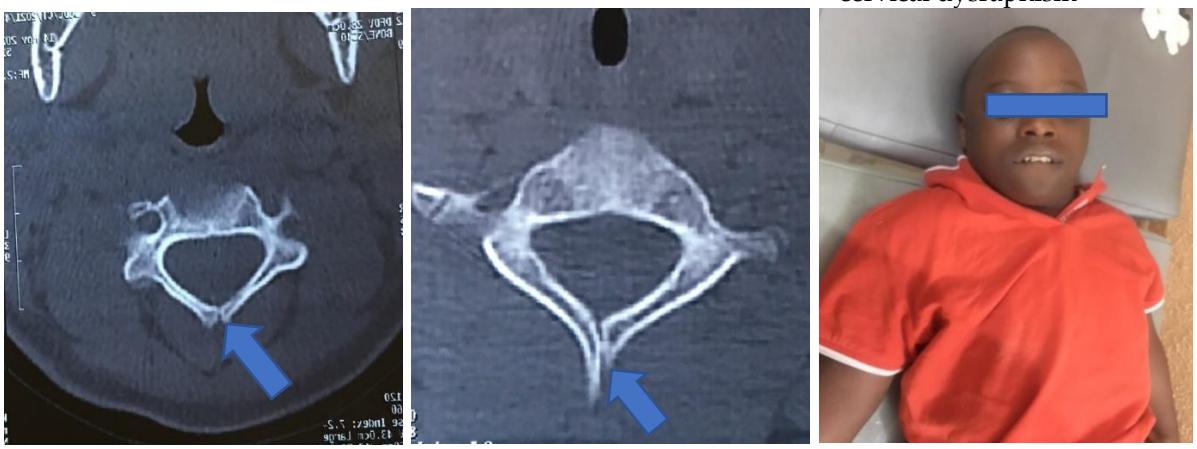


Figure 7. Cervical-3 dysraphism **Figure 8.** Cervical-7 dysraphism **Figure 9**. Down's Syndrome pupil with the two-level cervical dysraphism



The National Institute of Neurological Disorders and Stroke estimates that 10-20% of people may have spina bifida occulta.[2,3]

The regional distributions in this work showed that most were in the lumbar region (5 of 8), and the rest – cervical, thoracic and

sacral - had one dysraphism, each. These findings correlated closely with most publications which report the lumbar region with the modal frequency. Opsenak *et al*, in their publication reported that about 0-5% cases occur in the cervical spine and so, our 0.92% cervical occurrence correlated to this.[15]

Most of them were males, (male: female ratio of 3:1), correlating with the current knowledge in literature.[8,9,10,16] Also, this cohort of patients, being mostly of traumatic aetiologies, explained the finding of male preponderance. The tell-tale features were mostly abnormal hair growths on the skin over the lesion (7 of 8), while a lumbar dermal sinus was found on one subject.

The presenting diagnoses at the Emergency Room were quite wide, inclusive of intracranial tumours and degenerative spine disease. But, by far, the modal presentation (58.7% of the cases) was for head injury.

Historically, the prevalence of occulta is reported to be much higher than those for the other two types of overt dysraphism and estimates of its frequency have generally come from the incidental findings in spinal radiographs taken for reasons unrelated to the spine.[4,5]

Campbell, in his 2019 publication, reported that the incidence of spina bifida occulta, was much higher (10%) than that of the open dysraphisms, and Fidas *et al* even reported 17-30% incidence rate.[2,5] And so, in this work, the incidence of spina bifida occulta, which was 7.3%, closely compared with the incidence rates (5-30%) that had been reported by various scholars worldwide.[1,2,5,13,17] Perhaps, if this work was done as a community study, amongst the general population, the incidence may well be higher than our 7.3%.

There have been reports of an increase in the incidence of disc herniations with spina bifida occulta, especially in the presence of chromosomal abnormalities like Down's syndrome.[1,12,13] Our lone cervical spinal lesion was, interestingly, associated with Down's syndrome in a 9-year old male pupil who became unable to sit, stand or walk a few days after a classmate slapped him at the back of his head. Incidentally, his images revealed two bifid cervical spines, the 3rd and 7th; *see Figures 7, 8 & 9.*

Despite the challenges in the identification of this lesion in healthy individuals, the level of ignorance about the implication of the tell-tale signs, by itself, is a huge indictment on the knowledge, awareness and indices of suspicion of both our general population and healthcare givers.

Another aspect of the findings in this work was that the modal frequency for trauma (19.3%), was amongst the young males (73%). These findings are all in keeping with findings in literature. Aside the male gender preponderance amongst the 109, there were, also, more males affected by spina bifida occulta (6 of 8 or 75%) in this work.

Family or social correlations could not be traced along the history of any of the 8, despite the fact that several workers have reported a higher incidence in families where close relatives, especially from the mother's side, have had a history of spinal dysraphisms.

The tell-tale signs on the 8, were: dermal sinus 1 (0.92%), and abnormal growth of hair on the skin over the affect region 7 (6.42%), but only 6 presented with deficits. The 2 cases with sphincteric dysfunction included the one with a dermal sinus at the lumbar spine and sole patient with sacral dysraphism.

These tell-tale signs could be argued as red herrings, but when there is an established diagnosis of mild head injury, in a patient who suddenly develops an isolated foot drop, the chances of a correlation between the tell-tale signs and traumatic brain injury - which site is located far away from the lumbosacral spine, begins to look suspicious. And, when spinal x-rays reveal no radiological evidence of trauma- "spinal cord injury without radiological evidence of trauma" (SCIWORET), a closer scrutiny throws up a bifid spine.

Spinal magnetic resonance imaging, MRI, was done on 2 (1.83%), and computed tomography done on 2 (1.83%) other cases, because of inconclusive x-ray.

The radiological findings influenced the definitive management of these subjects, though their management fell outside the scope of this study.

CONCLUSION

Spina bifida occulta were detected amongst patients presenting with trauma (7.3%) affecting other parts of the body, outside the spine.

Trauma was the modal predisposing factor for the unmasking of this occult malformation and young males, constituted the modal population of victims.

All the cases of the occult dysraphism had tell-tale signs which had not, previously, been detected.

RECOMMENDATIONS

There is need to intensify the advocacy on the prevention of neural tube defects by using folic acid, in early pregnancy.

The challenge of neural tube defects, should be elevated to a public health policy.

The teaching curriculum and continuing professional education of healthcare professionals should continually reiterate the preventive measures of these defects.

There has, also, arisen the need for a multi-specialty National Association on Spinal Dysraphisms in Nigeria (NASD), as the fulcrum for this advocacy.

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