

Identification and Incidence of Spina Bifida during Gestation and its Pregnancy Outcomes

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ABSTRACT

Aim: To identify the incidence of spina bifida during gestation and its pregnancy outcomes.

Study design: Cohort study

Place and duration of study: Department of Radiology, Chandka Medical College Hospital, Larkana from 1st October 2020 to 30th September 2022

Methodology: One thousand pregnant women who were screened for the identification of spina bifida. Doppler based ultrasonography was performed in each patient. The fetus body was examined during sonography Doppler between 18-24 weeks. The body of the fetus was focused and visualized longitudinally in midsagittal section. Spinal cord was traced from neck to sacral, cervical vertebra and thorax. A tram track appearance was observed. Spinal nerve positioning was also observed. In spina bifida nerve tethering was seen. The primary outcomes in reference to baby weight and maturity were recorded while secondary outcomes of the fetuses in terms of any disability were followed up to 12 months of age.

Results: The mean age of the women was 25.6±5.6 years. The incidence of spina bifida was 1.1%. Within the primary outcomes observed birth weight ranged within 3.1-3.5kg. Around 81.81% cases were born full term while 18.18 were premature. On 06 months' follow-up of the spina bifida cases it was observed that 45.45% of new born suffered from skin problems as sores, calluses or and blisters while 36.36% had hydrocephalus with excessive CSF which needed monitoring. Mobility issue and disability was observed in 27.27% cases.

Conclusion: Spina bifida cases are identified successfully within gestational period. Prematurity can be significantly noticed in spina bifida cases. The lumbosacral and lumbar lesions are most common as well as skin conditions and hydrocephalus.

Key words: Incidence, Spina bifida, Gestation, Outcome

INTRODUCTION

Spina bifida is a term referring to a disease spectrum where caudal-neural tube fails to fuse developing embryo resulting into open, occult or dysraphism types. The open type is also known as spina bifida cystica refers to defects of caudal-neural tube where there is herniating in the spinal cord or and in meninges. It has two types as myelomeningocele and meningocele. Lesions are observed in the lumbar as well as sacral regions with severe deformity and cognitive dysfunction observed in higher lesions.^{1,2}

Within the aforementioned types the myelomeningocele is more common and lethal. It is related with infancy as well as childhood having downward medulla displacement, 4th ventricle and cerebellum through foramen magnum into cervical spinal-canal and also with the hydrocephalus condition. Cases which are affected require insertion of ventriculo peritoneal shunt mostly for the treatment of hydrocephalus. This condition can result onto lifelong mobility defects as well as urinary and bowel inconsistencies^{3,4}.

Spinal bifida occult is however a milder condition with majority being undetected. In this case the vertebral arch is not fused with the midline. However, the protruding of spinal cord is not presented with no external defects and lesion formation. The condition is benign with no neurological characteristics observed. Dysraphism includes the closed skin covered lesions with tethered cord. It can result in neurological as well and urological deformities⁵.

With the advancement in the technology of identification of spina bifida as well as the available surgical treatment technique there is an increased frequency of the cases surviving upto the age of fecundity with many cases able to conceive themselves⁶⁻¹⁰. The present study was designed to evaluate the identification of spina bifida during gestation and its outcomes in reference any disability.

The present study was conducted to assess the ultrasonographical accuracy in identifying the spina bifida cases during pregnancy. The results of this study will be one of the very few studies conducted in this region for assessing the health outcomes post.

MATERIALS AND METHODS

This cohort study was conducted in the Department of Radiology, Chandka Medical College Hospital, Larkana from 1st October 2020 to 30th September 2022. There were 1000 pregnant women who were screened for the identification of spina bifida. The sample size was generated through WHO sample size calculator where 95% CI and 80% power of test was taken for calculation. All pregnant women with no clinical history of previous spina bifida cases were included. Cases with high risk of abortion were excluded from the study. The study was ethically approved and the consent form was taken from each participant. A 3cc blood was withdrawn from each participant for measuring the alpha-fetoprotein (AFP) β-human chorionic gonadotrophin levels. Doppler based ultrasonography was performed in each patient. The fetus body was examined during sonography Doppler between 18-24 weeks. The body of the fetus was focused and visualized longitudinally in midsagittal section. Spinal cord was traced from neck to sacral, cervical vertebra and thorax. A tram track appearance was observed. Spinal nerve positioning was also observed. In spina bifida nerve tethering was seen. On the posterior valve signs of any protrusions were observed. In cases with spina bifida specifically in lumbar sacral region the skin protrusions were observed. The meninges protrusions and spinal protrusions were also assessed for identification of meningocele or myelomeningocele respectively. In fetal head signs for Arnold-Chiari malformation type 2 were assessed. In condition where positive sign was found the neural tube was downward stretched with posterior of the brain alteration and stretched banana/crescent shaped cerebellum, cisternamagna usually absence on midsagittal

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images. Lateral ventriclomegaly with excessive CSF. Change in head shape with lemon sign was also observed. Sagittal section of head was also observed with distance measured between brain stem and 4th ventricle as well distances from brain stem and occipital bone was measured. In normal condition it was maintained as 0.5:0.5 while in case of spina bifida the distance between brain stem and 4th ventricle with brain stem and occipital bone was 1:0.5. All demographic and clinical details were entered in the well-defined questionnaire. The primary outcomes in reference to baby weight and maturity were recorded while secondary outcomes of the fetuses in terms of any disability were followed up to 12 months of age. Data was analyzed using SPSS version 26.0 in terms of frequency and percentage.

RESULTS

The mean age of the women was 25.6±5.6 years with majority being within the age of 23-32 years. The mode of delivery for most of the women was through lower segment cesarean section (Table 1). Within the total cases there were 11 cases which were found positive for spina bifida among all cases (Fig. 1). There were 27.27% of the fetuses with myelomeningocele while 18.18% cases of each, lipomyelomeningocele, lipomeningocele and meningocele were also observed (Fig. 2).

The lesions were frequently observed in the lumbar and lumbosacral region with a percentage of 27.27% followed by lesions in sacral region respectively (Table 2). Within the primary outcomes observed there was 63.63% of the spina bifida fetus delivered as boys while 36.36% were girls. The birth weight ranged within 3.1-3.5kg. Around 81.81% cases were born full term while 18.18 were premature. On 6 months follow-up of the spina bifida cases it was observed that 45.45% of new born suffered from skin problems as sores, calluses or and blisters while 27.28% had hydrocephalus with excessive CSF which needed monitoring. Mobility issue and disability was observed in 18.18% cases (Table 3).

Table 1: Distribution of age and mode of delivery within cases (n=1000)

Variable	No.	%
Maternal age (years)		
17 – 22	170	17.0
23 – 32	510	51.0
33 – 42	320	32.0
Mode of delivery		
Vaginal	450	45.0
Lower segment section	550	55.0

Table 2: Lesion Level as observed in positive cases of spina bifida (n=11)

Lesions level	No.	%
Thoracic	1	9.09
Thoracolumbar	1	9.09
Lumbar	3	27.27
Lumbosacral	3	27.27
Sacral	2	18.18
Sacrococcygeal	1	9.09
Not available	-	-

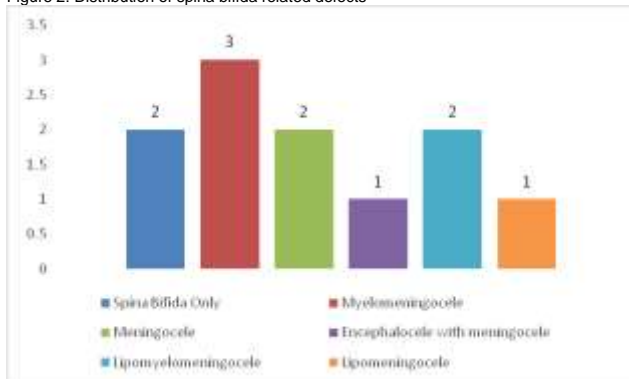
Table 3: Primary and secondary outcomes of spinal bifida born babies

Outcome	No.	%
Baby born		
Boy	7	63.64
Girl	4	36.36
Primary outcomes		
Birth weight (kg)		
Range	3.1-3.5	
Mean±SD	3.2±1.1	
Full Term Babies	9	81.82
Premature	2	18.18
Secondary outcomes		
Hydrocephalus	3	27.28
Mobility issues	2	18.18
Sores, calluses, blisters	5	45.45
Latex Allergy	1	9.09

Fig. 1: Frequency of spina bifida (Arrow)



Figure 2: Distribution of spina bifida related defects



DISCUSSION

There is a high frequency of women above the age of 32 years to have a risk of fetus spina bifida formation. The present study was conducted to assess the ultra-sonographical accuracy in identifying the spina bifida cases during pregnancy. This study identifies cases of spina bifida through high efficacy at ≤22 weeks of pregnancy. A cohort Danish study had reported the incidence of spinal bifida as 0.49%^{11,12}. In the current study the incidence was estimated as 1.1%. The prenatal detection rate prior to 22 weeks is measured to be higher than those examined post 22 weeks¹³.

Research has elaborated that “banana sign” of the cerebellum as well as “lemon sign” of the frontal skull are very important during identification of the spinal bifida.¹⁴ Closed spinal bifida impact on the carnial-structures is totally altered than is that of open spinal bifida. The sensitivity of the open spina bifida is 100% while that of closed spina bifida it was found around 92%¹⁵.

In developed countries cases of spina bifida with lethal deformities are lead to pregnancy termination; however such lethal deformities were not observed in this region. Moreover, termination of the pregnancy was considered ads the last option and was not considered by the pregnant women having fetus with spina bifida. In European countries the termination of spina bifida fetus ranged between 81-90%¹⁶⁻¹⁹.

Hydrocephalus is considered as a major concern in cases with spina bifida. In present study the most frequent presentation was of hydrocephalus cases. Studies have reported that Hydrocephalus is the major significant comorbidity in myelomeningocele from the neurosurgical perception. Clinically 75-80% of the patients having myelomeningocele required treatment with a shunt. However, within the recent years advancements in this field including intrauterine myelomeningocele-closure as well as ETV-CPC are decreasing this encumbrance²⁰.

CONCLUSION

Spina bifida cases are identified successfully within gestational period and are accompanied with normal birth weight. Prematurity can be significantly noticed in spina bifida cases. The lumbosacral

and lumbar lesions are most common. The hydrocephalus and Sores, calluses, blisters with mobility issues are frequently related with spina bifida cases.

Conflict of interest: Nil

REFERENCES

1. Detrait ER, et al. Human neural tube defects: developmental biology, epidemiology, and genetics. *Neurotoxicol Teratol* 2005; 27: 515–24.
2. Blasi I, et al. Myelomeningocele and pregnancy: a case report and review of the literature. *J Matern Fetal Neonatal Med* 2012;25: 1176–8.
3. Visconti D, et al. Sexuality, pre-conception counseling and urological management of pregnancy for young women with spina bifida. *Eur J Obstet Gynecol Reprod Biol* 2012; 163: 129–33.
4. Adzick NS, et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. *N Engl J Med* 2011; 364: 993–1004.
5. Office for National Statistics (UK). Trends in neural tube defects. *Health Statistics Quarterly* 2001; 10: 9.
6. Oakeshott P, et al. Expectation of life and unexpected death in open spina bifida: a 40-year complete, non-selective, longitudinal cohort study. *Dev Med Child Neurol* 2010; 52: 749–53.
7. Mitchell LE, et al. Spina bifida. *Lancet* 2004; 364: 1885–95.
8. Fimmel R, et al. Does prenatal screening for 5,10-methylenetetrahydrofolate reductase (MTHFR) mutations in high-risk neural tube defect pregnancies make sense? *Genet Test* 2002; 6: 47–52.
9. MRC Vitamin Study Research Group. Prevention of neural tube defects: results of the Medical Research Council Vitamin Study. *Lancet* 1991; 338: 131–7.
10. Moore LL, et al. Folate intake and the risk of neural tube defects: an estimation of dose-response. *Epidemiology* 2003; 14: 200–205.
11. McComb JG. A practical clinical classification of spinal neural tube defects. *Child's Nervous System* 2015; 31(10): 1641–57.
12. Hyett JA. The Danish Fetal Medicine Database: Revealing the fruits of collaborative research. *Acta Obstetrica et Gynecologica Scandinavica* 2015; 94(6): 561-2.
13. EUROCAT. Detailed Congenital Anomaly Coding Guidelines 2013 [cited 2017 December 16], <http://www.eurocat-network.eu/content/EUROCAT-Guide-1.4-Section-3.5.pdf>.
14. Alriksson-Schmidt, M. Arner, L. Westbom et al. A combined surveillance program and quality register improves management of childhood disability. *Disability Rehab* 2017; 39(8): 830–36.
15. Ghi T, Pilu G, Falco P, et al. Prenatal diagnosis of open and closed spina bifida. *Ultrasound Obstet Gynecol* 2006; 28(7): 899–903.
16. Lennon CA, Gray DL. Sensitivity and specificity of ultrasound for the detection of neural tube and ventral wall defects in a high-risk population. *Obstet Gynecol* 1999; 94(4): 562-6.
17. Fleurke-Rozema JH, Vogel TA, Voskamp BJ, et al. Impact of introduction of mid-trimester scan on pregnancy outcome of open spina bifida in the Netherlands. *Ultrasound Obstet Gynecol* 2014; 43(5): 553-6.
18. EUROCAT. Prenatal Detection Rates 2017 [cited 2017 2 December], [http://www.eurocat-network.eu/prenatalscreeninganddiagnosis/prenataldetection\(pd\)rates](http://www.eurocat-network.eu/prenatalscreeninganddiagnosis/prenataldetection(pd)rates).
19. EUROCAT. Prevalence Tables 2015 [cited 2017 2 December], <http://www.eurocat-network.eu/accessprevalencedata/prevalencetables>.
20. Blount JP, Maleknia P, Hopson BD, Rocque BG, Oakes WJ. Hydrocephalus in spina bifida. *Neurol India* 2021; 69(Supplement):S367-71.