International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor (2018): 7.426

Myelomeningocele at a Tertiary Care Hospital - Social and Medical Factors, Dietary Habits Influence on Occurrence

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Abstract: ***Introduction: Folic acid deficiency in antenatal period is a well known etiological factor for spinal dysrapahism. Still many regions of Rural India are not aware of the importance of folic acid supplementation and antenatal screening for congenital anomalies. Objectives: To study the folic acid supplementation and antenatal ultrasonography screening to rule out structural anomalies in mothers of children with spinal dysraphism. Also to know various social factors contributing to the delayed detection and occurrence of spinal dysraphism. Materials and Methods: This is a retrospective study on the mothers of children with Spinal Dysraphism from rural areas of North Costal Andhra Pradesh. Results and Conclusion: Most of them were unaware of folic acid supplementation, its advantages, usage and effects of its deficiency. None of them started folic acid before planning to conceive. Only few of the cases were detected in antenatal ultra sonogram screening.***

Keywords: Folic acid supplementation, Rural, Radiologist, Spinal dysraphism , (TIFFA) targeted imaging for foetal anomalies scan

1. Introduction

Annually 3-4 lakh infants are born with spina bifida and anencephaly worldwide 1. In India each year 1-3 of 1000 live births are affected with spinal dysraphism 2. Children develop paraplegia, sphincter disturbances, CSF leaks, hydrocephalus due to ACM and learning disabilities and delayed milestones with significant physical disability, psychosocial maladjustments and increased financial burden to the family[3,4]. Folic acid deficiency is a well known etiological factor for neural tube defects. Folic acid supplementation has proved beyond doubt as a preventive measure for spinal dysraphism[5]. Screening with TIFFA scan can detect NTD as early as 18-20 weeks[6]. When TIFFA scan is done by a radiologist patients can elect for termination of pregnancy at the earliest.

Consanguinity, other social factors contributing to the ignorance of people about preventive measures have been playing its role.

2. Objectives

The following are the objectives of the study among mothers of spinal dysraphism children in the Rural areas of the three districts of Srikakulam, Visakhapatnam and Vizianagaram of North East Costal belt of Andhra Pradesh state. 1. To study the awareness of folic acid supplementation and the start of folic acid supplementation in relation to gestation of pregnancy. 2. To study the social factors like mother's consanguinity, literacy and tribal (low socio economic status) on the general awareness of the disease. To study antenatal ultrasonography screening - scans during pregnancy, gestational age at which first scan was done, scan done by radiologist or inexperienced gynaecologist, cases identified by ultrasonography.

3. Materials and Methods

It is a retrospective study done at Neurosurgery Department, King George Hospital, Visakhapatnam from the period between Jan 2016 and december 2018.One hundred and eight mothers of children with spinal dysraphism , who belong to rural areas of three districts of Srikakulam, Visakhapatnam and Vizianagaram of North east Costal Andhra Pradesh state are included in the study. The literacy, consanguinity, and social status of mothers were known, enquired about the awareness of folic acid supplementation and the gestational age at which they started the supplementation. They were also questioned about the antenatal ultrasonography screening done to rule out structural abnormalities. Also enquired if the scans were done by radiologist or inexperienced gynaecologist which would influence the probability of identification and prevention of these spinal abnormalities.

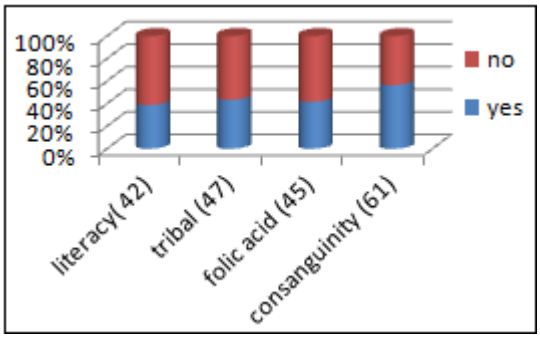

Figure 1: Results

Of the total 108 cases (FIGURE 1) in the study,61 (56.4%) were products of consanguineous marriage,46% were of 1st degree relative, coming from a low socioeconomic background.

Almost in 43% (47) of the cases, were hailing from tribal areas around the above mentioned areas around Visakhapatnam with no proper medical attendents to guide the pregnancy period. Of these most of the mothers were deprived of the medical facilities with nutrition and regular check ups

Only 38.8% (42) of the mothers were found literates in our study making it a significant factor in general awareness among the public in terms of seeking timely medical help and monitoring.

Of the total 108 cases in the study only 41%(45) had taken folic acid from the 6 th or 7 th month onwards only after they had visited the hospital with complaints of weakness and anaemia for which they were prescribed folic acid along with iron supplementation. None of the mothers started folic acid since conception. This shows there has been gross unawereness even among the medical teams in proper evaluation and timely medication to the patients.

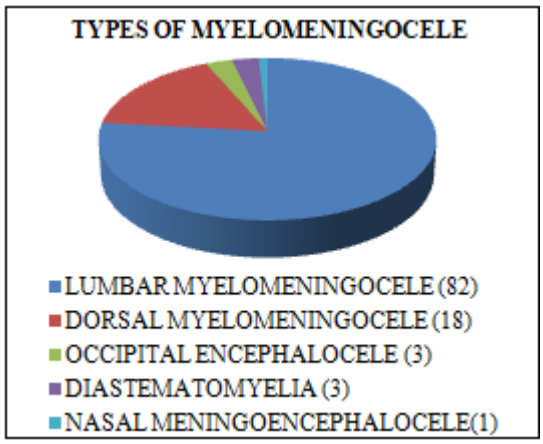

Figure 2

In the study of three years from 2016 to 2018,among the meningomyelocele cases(FIGURE 2),82 were lumbar myelomeningocele making it the most common presentation followed by dorsal meningomyelocele (18),occipital encephalocele(3),and diastematomylia. There were no cases with combined or multiple lesions.

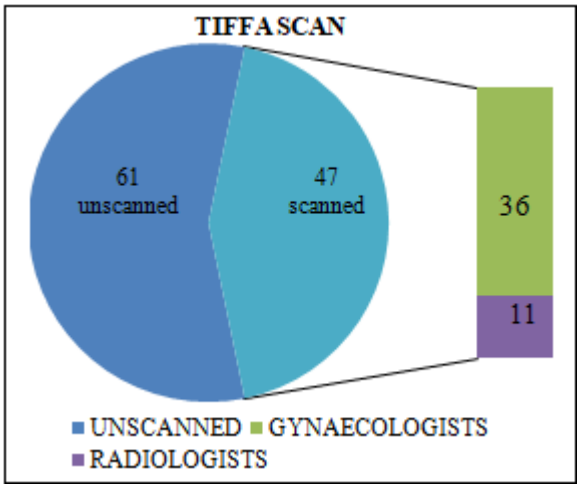

Figure 3

In the study over three years, it has been echoing that sonographic screening is crippled in early detection of these spinal dysraphism. Only (FIGURE 3) 47 mothers were scanned for foetal parameters of baby in the womb and among those, 36 mothers underwent ultrasound screening mostly by gynaecologists.

So radiologically in experienced gynaecologists weren't aware of the findings or lacked the intent to look for spinal deformities.

However, 11 mothers were screened by radiologists and 9 detected spinal meningomyelocele although at around 7th month which were not of much benefit.

Table 1

Associated foetal anomalies with myelomeningocele	
Hydrocephalus	68
Anencephaly	0
Paraplegia	49
Limb deformities: Club foot, valgus deformities	32

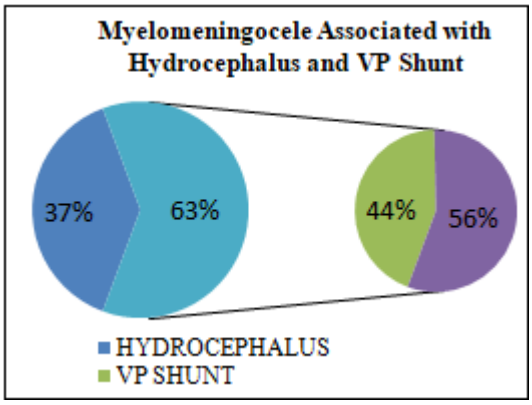

Figure 4

63% of the patients in the study had congenital hydrocephalus along with spinal meningomyelocele, among them 44 % of the patients underwent ventriculo peritoneal shunting either preoperatively or post operatively of myelominingocele repair (Figure 4).

There comes the role of expert radiologist or experienced gynaecologist who can detect these spinal dysraphism along with congenital hydrocephalus and guide the parents into decision of continuing pregnancy.

Also depicts the increase in burden on the parents especially from the low socioeconomic background with prolonged hospital stay and perioperative expenses and mental toil.

4. Discussion

Annually about 2,500 infants are born with the neural tube defects, spina bifida and anencephaly. All infants with anencephaly die shortly after birth, whereas the majority of babies born with spina bifida grow to adulthood with, in severe cases, paralysis and varying degrees of bowel and bladder incontinence.

The offspring of consanguineous unions are more likely to inherit homozygous alleles than are the offspring of

unrelated parents. In consanguineous marriages the inbreeding coefficient (F) equals or is higher than 0.0156, where (F) is representative of a measure of the fraction of loci at which the offspring of a consanguineous union is expected to inherit identical gene copies from each parent[18] Consanguinity allows clustering of susceptible genes,expression of which could probably contribute to development of neural tube defects.

The evidence that consumption of folic acid, one of the B vitamins, before conception and during early pregnancy (the periconceptional period) can reduce the number of NTDs has been accumulating for several years[5,6].

The active form of folic acid is the reduced form called tetrahydrofolate (THF).The prime function of folate is to provide one-carbon moieties for the synthesis of three of the four bases of DNA, guanine, adenine and thymine. Several dietary sources are rich in natural folate, such as fruits, green leafy vegetables, i.e. Spinach, Brussel sprouts and broccoli, oranges, beans, yeast and liver. After oral ingestion, following metabolism, it is fully reduced to tetrahydrofolate (THF) by the enzyme tetrahydrofolate reductase. THF can be transported directly into the portal circulation or be converted to THF-polyglutamate stores or to 5-methyl THF monoglutamate, the predominant form of folate in serum and tissues

All women of childbearing age who are planning pregnancy should consume 0.4 mg of folic acid per day for the purpose of reducing their risk of having a pregnancy affected with spina bifida or other NTDs. Because the effects of higher intakes are not well known, care should be taken to keep total folate consumption at less than 1 mg per day, except under the supervision of a physician. Women who have had a prior NTD-affected pregnancy are at high risk of having a subsequent affected pregnancy. When these women are planning to become pregnant, they should consult their physicians for advice.

Sonographic diagnostic accuracy for open neural tube defects has improved dramatically in recent years. Enhanced image resolution has allowed localization of the origin of the echoes from the immature fetal spine and identification of significant cranial and intracranial anatomic changes in the presence of open spina bifida.

Recent investigations indicate that the sensitivity of sonography for open spina bifida is 95% when the examination is a targeted scan by an experienced radiologist.

Sonographic evaluation of the fetal spine depends on visualisation of the ossification centres within the fetal vertebrae. In the transverse plane, all three ossification centres should be visualised, and the centres of the neural arches should be parallel or converging. the parallel configuration is particularly noticeable when the foetus is in decubitus position with respect to the transducer[15]. In the longitudinal plane, the spine has a “rail-road track” appearance, with gradual widening toward the fetal head and gradual tapering in the sacrum. The distal part of the spine may not be ossified in healthy foetuses before 22 weeks of gestation, and in such cases imaging may be repeated if the foetus is at high risk for a neural tube defect.

Transvaginal or transperineal imaging in the second trimester can be extremely helpful in visualising the distal spine of the fetus in a persistent breech presentation.

Prenatal detection of spina bifida has become very important, as termination of pregnancy before fetal viability is elected by upto 80% of families. If the pregnancy is continued, detection may affect the delivery location and method. The degree of handicap and the survival rate depend on the level of the spinal segments involved, the severity of the lesion, the treatment program and the associated abnormalities.

5. Conclusion

With the study results,its clear that consanguinity ,literacy, social status have been playing major roles in public ignorance towards the medical facilities and proper health care during pregnancy. Out of this unawareness, folic acid supplementation which has been proven to have decreased the cases of spinal dyraphism has not been into total effect in the tribal,low socioeconomic section. It also brings out the great need for the screening tests, the ultrasonographic detection at an early stage by an expert radiologist or atleast gynaecologists with experience and training to take up the role, by genetic counselling help the parents make decisions properly in time. It is upto the government and the health care system to spread awareness among these sections and provide them the necessary medical aid in early detection ,prevention and management of these congenital diseases.

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