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An Overview On Spina Bifida

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***Abstract*: Spina bifida is a congenital malformation in which the spinal column is split (bifid) as a result of failed closure of the embryonic neural tube, during the fourth week post- fertilization. It is the second leading cause of birth defects after the congenital heart defects. This may result in lower limb weakness or paralysis that hampers or prevents walking and lack of sensation that enhances the risk of pressure sores. In this review paper, we discuss the phenotypes seen in humans as described by both surgoens and embryologists and contrast it to the leading animal model, the mouse. Thus, here we will come to know about spina bifida in all its forms both aperta and occulta and the measure of deterioration arising from caudal neural tube defects, known as spina bifida, must be determined by the level of lesion both in mouse and in man.**

***Keywords*: Lesion, Meningocele, Myelomeningocele, Spina bifida aperta, Spina bifida occulta.**

1. Introduction

Spina bifida is the most common birth defect affecting the central nervous system and is often characterized as the most complex birth defect compatible with survival. It often causes paralysis of the lower limbs and sometimes learning difficulties. It occurs when the spine and spinal cord Do not form properly. The study of this condition is extremely relevant in that even in the 20 years, since the discovery of the benefits of folic acid this condition is highly prevalent around the world and it's occurrence does not seem to be decrease. This review paper intends to research on spina bifida to increase the awareness and treatment strategies and to compare and contrast spina bifida in humans and spina bifida in the mouse.

2. Epidemiology

The prevalence of spina bifida is large because the prevalence of neural tube defects is declining in North America and western Europe because of dietary fortification and also because of advanced prenatal diagnosis that is leading to more elective terminations. The parents infrequently terminated fetuses with non-lethal defects, with spina bifida showing a 7-10 termination. Despite these findings, spina bifida is not giving away as a common congenital birth defect. Additional births involving spina bifida have been documented in families with the previous births who were taking dietary supplements not only is spina bifida not going to disappear but also there are still several thousand pregnancies involving spina bifida live in the United States [1].There is an urgent need to develop a comprehensive research program focusing on people with spina bifida, which will also have important implications for enhanced scientific understanding and treatment of other neuro developmental disabilities. It has an incidence generally around 0.5 per 1,000 births, although higher frequencies have been reported. The studies in parts of Asia have also shown a lower occurrence of spina bifida that that of uk.

3. Etiology

The etiology of spina bifida is heterogeneous [2]-[5] most nonsyndromic spina bifida6 is thought to be of multifactorial origin with influence of both genetic and environmental factors. Among the environmental factors associated with increased risk of spina bifida are incresed pregnancy weight [7]-[13], maternal smoking [14], [15], drug intake specifically of antiepileptic drugs [16], [17] and maternal illness such as diabetes [18], [19] and hyperthermia [20].

4. Pathogenesis

The spina bifida occulta is the second major form of NTDS, where the site of the lesion is not left exposed. The spinal dorsal dermal sinus tract ranging phenotypically from dysplastic skin, tuft of hair and vestigial tail as well as other forms of spinal dysraphism where the vertebrae develop abnormally leading to neural arches and can result in pain, Weakness. Myelomeningocele [21], [22] is usually associated with a type 2 chiari hind brain malformation. It is the downward displacement of the cerebellar vermis into the cervical vertebral canal. This malformation causes elongation of brain stem and obliteration of the fourth ventricle leading to obstruction of cerebro-spinal fluid. It is often symptomatic and is diagnosed prenatally with ultrafast fetal magnetic resonance imaging. (MRI) [23], [24]. Meningocele is often described as a less severe variant of myelomeningocele in which the spinal cord is not found in sac and is described by embryologists to be absent of neural matter in its herniated sac. The status of meningocele being an open or closed defect is still debatable in terms of embryogenesis. Spina bifida aperta, sometimes referred to as spina bifida cystica, is Usually visible at birth as an exposed neural tissue with or without a protruding sac at the site of the

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lesion. It protrudes from the spinal canal into a fluid – filled sac resulting from incomplete closure of the primary neural tube.

5. Spina bifida in Humans

Development of the central nervous system including the brain and spinal cord is a complex process beginning with a flat sheet of cells which undergoes sequential thickening, elevation, mediolateral convergence accompanied by rostrocaudal extension and finally adhesion to form the neural tube which is the precursor of the brain and the spinal cord. NTDS can occur in two major forms: spina bifida aperta, which is the open-lesion NTD, and the closed-lesion NTD, more commonly known as SB occulta.

6. Spina bifida in Mouse

There exist more than 250 mouse models with neural tube defects of which 74 are of spina bifida. It does not mean that the studies on the [25] structural changes afforded by mouse model cannot be used as a tool to understand human spina bifida. In vertebrates, the development of the CNS starts with the formation of the neural plate on the dorsal surface of the embryo during late gastrulation [26], [27]. Primary neurulation is responsible for formation of the neural tube throughout the brain and spinal cord rostral to mid- sacral level [28]. Morphologically [29]-[32], the mouse neural tube undergoes distinct structural changes prior to it's closure. In mouse and humans spina bifida occulta has largely been described as a result of failure of secondary neurulation. Secondary neurulation in the mouse is described as occurring at the sacral level.

7. Haploin Sufficiency in Mouse and Man

The occurrence of spina bifida in genes acting in an additive or the subtractive manner is almost unknown. There are 5 studies in the mouse, which have demonstrated spina bifida and the interaction of the involved genes mechanistically. These include Lrp6 and Wnt5a [33], Zacl and Suz12 [34], Hira and Pax3 [35], Rybp encompassing Ringl, Yypl [36] and haploin sufficiency of the components in the primary cilium of hedgebog pathway [37].

8. Treatment and Management

Management of patients were sometimes denied Treatment based on the severity of their condition. The management of mmc traditionally involves surgery within 48hours of birth. The child's back is closed to minimize the risk of ascending infection that can result in meningitis.

A. Diagnosis and Screening

prenatal diagnosis first became possible in early 1970s [38], [39], with the finding of an elevated concentration of alphafeto protein in amniotic fluid samples from pregnancies with anencephaly. Biochemical Screening for mmc [40] is becoming redundant as ultrasound offers greater sensitivity and specificity.

B. Sonographic Diagnosis

The fetal spine can be examined by ultrasonography in the sagittal, axial and coronal planes from late first trimester onwards providing the principle and most accurate mode of prenatal diagnosis.

C. Post-natal Surgery and Management

Neonates with spina bifida are best managed following baseline imaging studies of the central nervous system and subsequent serial head measurements to assess the velocity of head growth and need for shunting [41] medical management of the individuals with spina bifida is best provided through regular assessments by multidisciplinary team. Additional issues that may need to be addressed by team include neurobehavioral development, weight maintenance and skin care.

D. Fetal surgery

The rationale for fetal surgery [42] is that damage to the exposed spinal cord is progressive during gestation, hence early repair of the lesion, in utero, may prevent continuing damage and improve clinical outcome. Skin flaps are widely mobilized and closed to complete the repair although, when the skin cannot be closed primarily [43].

9. Prevention and Detection

Dietary factors including water chlorination [44], inositol intake [45], simple sugar intake [46] and intake of trace elements and other micronutrients [47].

- Elevated levels of maternal serum alpha-fetoprotien are usually indicative of spina bifida aperta [48], [49].
- Screening obstetrical ultrasonography is the initial routine method for the detection of NTDS during pregnancy.

10. Conclusion

This review paper aims to probe spina bifida, the surviving form of neural tube defects, closely and to analyze the relationship of what can be learnt from a mouse model of spina bifida and to use that knowledge in order to understand with regard to the human form.

The article addresses the etiologies of spina bifida, brain/behavior relationships, specific medical issues, and contextual influences of family, school, society, and health care delivery, the undercurrent theme is that more research is needed—research that multi-site, collaborative, encompasses the lifespan, and incorporates functional assessment, measures of activities, societal participation, and quality of life of persons with spina bifida as important components.

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