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EPIDEMIOLOGY OF NEURAL TUBE DEFECTS IN KING SALMAN ARMED FORCED HOSPITAL OF TABUK REGION, SAUDI ARABIA BETWEEN 2012-2017

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Abstract:

Retrospective study is conducted to analyze the Neural Tube Defects (NTDs) in King Salman Armed Forced Hospital of Tabuk Region, Saudi Arabia between 2012-2017. The overall number of cases was 49. The meningomyelocele (MMC) with hydrocephalus (HC) is the most common type of (NTDs) in this study with 55.1% incidence rate. Congenital Heart Diseases are the most common associated anomaly with (NTDs) from the systemic organs side, with 10.2% incidence rate. The study showed that 16.3% of the cases developed a permanent neurological defect and disability, mostly paraplegia 10.2 %, lower limbs weakness 4.1%.

The data have shows that this surgical intervention was complicated with Meningitis in 8.2% of the cases, and 4% complicated with surgical site infections, 50% of all post-operative meningitis occurs if both repair and ventriculoperitoneal (VP-Shunt) done together.

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INTRODUCTION:

Neural tube defects (NTDs) are severe birth defects of the central nervous system that originate during embryonic development when the neural tube fails to close completely. Human (NTDs) are multifactorial, with contributions from both genetic and environmental factors.[1]

Individuals with spina bifida often require walking aids and wheelchairs for functional mobility. In the United States of America (USA)in any given year, on average, 33% of individuals with spina bifida made claims from Medicaid for some type of mobility-related assistive technology, including wheelchair-related costs, orthotics and prosthetics, ambulatory aids, and communication and hearing aids. Annually, these claims accounted for €297,704 (price year not reported), which represents approximately 3.3% of all reimbursement by Medicaid for all medical care for these individuals. In all the countries where cost–benefit of folic acid for the prevention of (NTDs) was evaluated, several millions to hundreds of millions of Euros (or dollars) of net benefit or cost savings were estimated. These results strongly support the continuation of folic acid for the prevention of (NTDs), especially in countries with high prevalence of (NTDs). [2]

RESULTS:

-The study are composed 49 patients in which they are diagnosed and managed in King Salman Armed Forced Hospital (KSAFH) of Tabuk Region, (KSA) ,male and female, the 33 patients of them are males (67.3%), and 16 females (32.7%), and (4.1%) of them have the family history of (NTDs).

-The study shows that the consanguinity is higher concern among those patients and their families, where there are 55.1% of the cases are positive consanguinity relation, in compared with 6.1% who are documented as negative consanguinity, taking consideration of 36.7% who are not documented their relationship in the medical records.

- The study reveals that the meningomyelocele (MMC) with hydrocephalus (HC) is the most common type of (NTDs) in this study with 55.1% incidence rate, followed by congenital hydrocephalus with 34.7%.

-The study shows that 49% of the patients are free from associated congenital anomalies in terms of systemic diseases or neurological related diseases.

- Regarding the association between (NTDs) and the neurological problems, the Seizure was the most common associated with 14.3 %, followed by Scoliosis 10.2%, and the other associated neurological peoblems with (NTDs) is show in (Graph 1-2)

-From the surgical intervention side, the study reveals that 95.5% of the all patients was proceeded for the surgery, where 46.9% have done both the repair and Ventriculoperitoneal Shunt (VP Shunt), a 32.7% proceed for (VP Shunt) only, and 16.3% proceed for the repair only.

- The data have shows that this surgical intervention was complicated with Meningitis in 8.2% of the cases, and 4% complicated with surgical site infections.

- Regardless the limitations in the data, the results of folic acid supplements, Genetic with Chromosomal Counseling, prenatal procedures and medications are arranged in the (Table 1-1)

- The study showed that 16.3% of the cases developed a permenant neurological defect and disability, mostly paraplegia 10.2 %, lower limbs weakness 4.1%.

- The study reveals that 42.85% have a systemic congenital anomalies, 61.9% of them are male, and the other 38.1% of them are female.

- Overall, the Congenital Heart Diseases are the most common associated issue with (NTDs) from the systemic organs side, with 10.2% incidence, followed by Neurogenic bladder with 4.1%, and the other associated anomalies with (NTDs) is show in (Graph 1-1)

- 80% of all (NTDs) with Congenital Heart Diseases are consanguineous married.

- 50% of all post-operative meningitis occurs if both repair and (VP Shunt) done together.

- The study shows that only 25.9% of those consanguineous married who are done a Genetic and Chromosomal Counselling prior to conceive.

DISCUSSION:

The Neural Tube Defects (NTDs) are classified to four main subtypes which is : Hydranencephaly, Holoprosencephaly, Microcephaly, Macroencephaly.

Hydrocephalus : A post-neurulation defect. Total or near-total absence of the cerebrum (small bands of

cerebrum may be consistent with the diagnosis), with intact cranial vault and meninges, the intracranial cavity being filled with CSF. There is usually progressive macrocrania, but head size may be normal (especially at birth), and, occasionally, microcephaly may occur. Facial dysmorphism is rare.

May be due to a variety of causes, the most commonly cited is bilateral Internal Carotid Artery (ICA)

infarcts (which results in absence of brain tissue supplied by the anterior and middle cerebral arteries with preservation in the distribution of the Posterior cerebral artery). May also be due to infection (congenital or neonatal herpes, toxoplasmosis).

Less affected infants may appear normal at birth, but are often hyperirritable and retain primitive reflexes (Moro, grasp, and stepping reflex) beyond 6 mo. They rarely progress beyond spontaneous vowel production and social smiling. Seizures are common.

Holoprosencephaly: Failure of the telencephalic vesicle to cleave into two cerebral hemispheres. The degree of cleavage failure ranges from the severe alobar (single ventricle, no interhemispheric fissure) to semilobar and lobar (less severe malformations). The olfactory bulbs are usually small and the cingulate gyrus remains fused. Median faciocerebral dysplasia is common, and the degree of severity parallels the extent of the cleavage failure. 80%are associated with trisomy (primarily trisomy 13, and to a lesser extent trisomy 18). Survival beyond infancy is uncommon, most survivors are severely retarded, a minority are able to function in society. Some develop shunt dependent hydrocephalus. The risk of holoprosencephaly is increased in subsequent pregnancies of the same couple.

Microcephaly : Head circumference more than 2 standard deviations below the mean for sex and gestational age. Terms that are sometimes used synonymously: microcrania, microcephalus. Not a single entity, many of the conditions may be associated with microcephaly. It may also result from maternal cocaine abuse.It is important to differentiate microcephaly from a small skull resulting from craniosynostosis in which surgical treatment may provide opportunity for improved cerebral development.

Macroencephaly :Enlarged brain which may be due to, hypertrophy of gray matter alone, gray and white matter, presence of additional structures (glial overgrowth, diffusegliomas, heterotopias, metabolic storage diseases).

Conditions in which macrocephaly may be seen include:

-Neurocutaneous syndromes (especially neurofibromatosis) [1]

-Megalencephaly-capillary malformation syndrome (MCAP): an overgrowth syndrome with megalencephaly (often with hydrocephalus, Chiari malformation, polymicrogyria and seizures), and capillary malformations in the skin (usually on the face).[3]

The risk factors for Neural Tube Defects :

1- Lack of prenatal folic acid: early administration of folic (0.4 mg/d if no history of neural tube defects; 4 mg/d in a carrier or with previous child with (NTDs)[4]

2- Folate antagonists (e.g. carbamazepine) doubles the incidence of MM, and use of valproic acid during pregnancy is associated with a 1–2%risk of (NTDs)[5]

3- Obesity (before and during pregnancy) increases the risk of (NTDs)[6][7]

4- Maternal cocaine abuse may increase the risk of microcephaly, disorders of neuronal migration, neuronal di erentiation and myelination. [8]

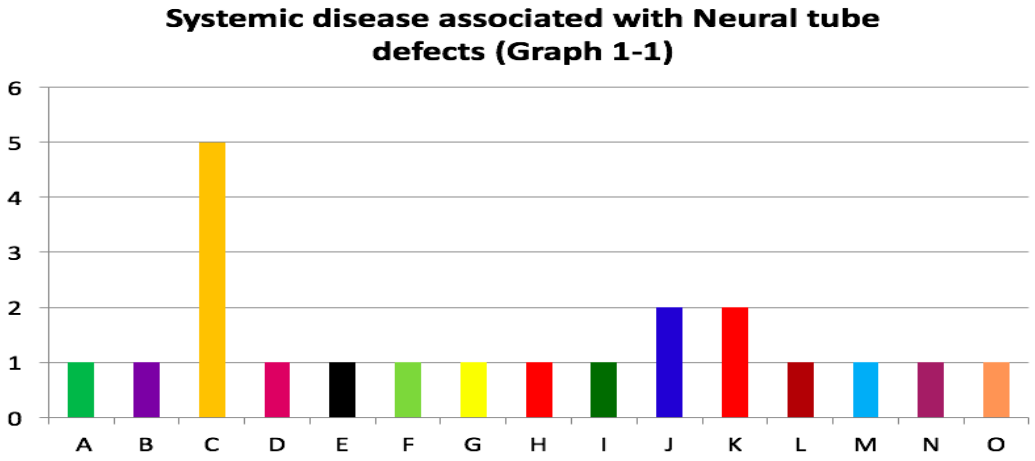
Prenatal procedures that can be done to detect the (NTDs) :

1- Serum alpha-fetoprotein (AFP) : A high maternal serum AFP (≥2 multiples of the median for the appropriate week of gestation) between 15–20 weeks gestation carries a relative risk of 224 for neural tube defects, and an abnormal value (high or low) was associated with 34%of all major congenital defects.[9]

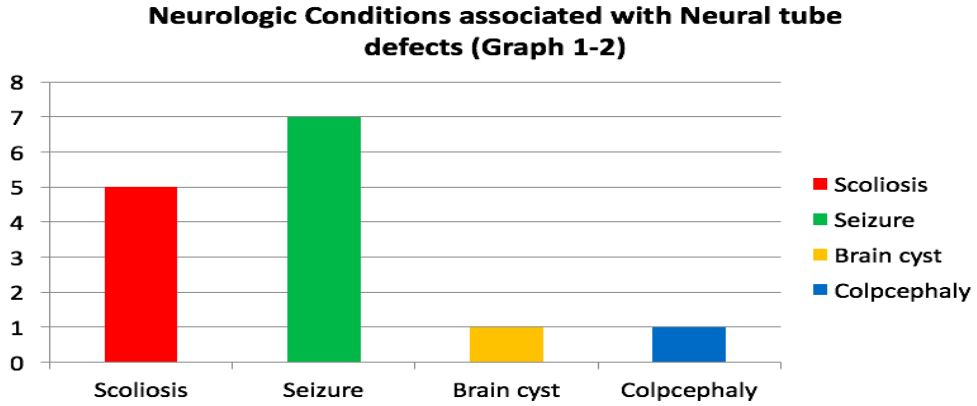
2-Ultrasound : Prenatal ultrasound will detect 90–95%of cases of spina bifida, and thus in cases of elevated AFP, it can help differentiate NTDs from non-neurologic causes of elevated AFP (e.g. omphalocele), and can help to more accurately estimate gestational age.

3- Amniocentesis: For pregnancies subsequent to a myelomeningocele, if prenatal ultrasound does not show spinal dysraphism, then amniocentesis is recommended (even if abortion is not considered, it may allow for optimal post- partum care if

myelomeningoceleis diagnosed). Amniotic fluid AFP levels are elevated with open neural tube defects, with a peak between weeks 13–15 of pregnancy.



A) Developmental Delay
B) Renal Anomalies
C) Congenital Heart Diseases
D) Achondroplasia
E) Congenital Toxoplasmosis
F) Neurogenic Bladder
G) MI + Thrompocytopenia
H) Perforated ear + Paralytic Talipes
I) Heart Disease + Atonic Anus
J) Club Foot + Neurogenic Bladder
K) Heart Disease+ Neurogenic Bladder
L) Pulmonary stenosis + Neurogenic Bladder + HTN
M) Neurogenic Bladder + Single Umbilical Artery + Dysmorphic Features
N) Heart Disease + Hip Dislocation



The measure that help to diagnose NTDs and prevent it (Table 1-1).

	Yes	No	Unknown
Folic acid supplements	**6.1**	**4.1**	**89.8**
Prenatal Procedures	**12.2**	**87.8**	**100**
Prenatal medications	**2**	**6.1**	**91.8**
Genetic and chromosomal counseling	**18.4**	**2**	**79.6**

CONCLUSION:

Neural tube defects are a common and preventable condition. The population awareness remains a major public health challenge to prevent, anticipate, early diagnosis, and neonatal neurosurgical intervention as the most important factors in reducing immediate morbidity and mortality. In our study, we find there is deficient of awareness regarding the benefits of the folic acid during pregnancy, deficient awareness regarding genetic and chromosomal Counseling.

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