

A STILL BORN BABY WITH CRANIAL VAULT DEFECTS

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CASE HISTORY

A 40-year-old-mother presents to the antenatal clinic with uterus size larger than gestational age. The gestational age is 32 weeks but uterine size corresponds to term size. She comes from a rural village without any early antenatal booking. Ultrasound confirmed polyhydramnios and fetal abnormality. She has no significant past medical history. Two weeks later she delivers her baby via spontaneous vaginal delivery. The baby is still born (Figure 1).

QUESTION

1. Describe the abnormalities present in the newborn and state the diagnosis.
2. What is the pathogenesis of this condition?
3. What are the risk factors for this condition?
4. What are the preventive measures which can be implemented prior to pregnancy?



(Original photo taken by the authors)

ANSWER:

1. The photo shows a developmental defect of the baby's skull. The brain and cranial vault are severely deformed. The fore brain (Cerebrum) appears hypoplastic and malformed and it is exposed without the covering of the cranial vault. The diagnosis is anencephaly. The reported prevalence of anencephaly was 1.29 per 1000 births in Malaysia which is not different from that reported in developed countries.¹
2. Anencephaly is a form of neural tube defects which usually occurred during the 3rd to 4th week of embryo development. It is caused by the failure of closure of the neural tube at the cranial end of the embryo.² It is usually multifactorial but genetic factors such as MTHFR gene (5,10-methylenetetrahydrofolate reductase) and VANGL1 (Vang-like 1) protein have been known to be associated with this condition.^{3,4}
3. Among the common risk factors for anencephaly are: maternal diabetes mellitus, previous history and family history of neural tube defect, folate deficiency and valproic acid treatment of epilepsy in pregnancy.^{1,4,5}
4. There is enough evidence that adequate intake of folic acid prior to pregnancy can minimize the risk of anencephaly. Women of reproductive age are recommended to consume 0.4 mg folic acid daily.^{1,5,6} The dosage should be increased in pregnant mother who are in the high risk group especially those with past history and family history of anencephaly.

Folic acid is best to be consumed three months prior to conception and continue through first trimester. Study has shown that women who consume adequate folic acid has reduce incidence of open neural tube defect up to 25%.^{5,6}

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Older age and longer duration of previous quit attempts predict successful smoking cessation

Wee LH, Shahab L, Bulgiba A, *et al*. Stop smoking clinics in Malaysia: characteristics of attendees and predictors of success. *Addict Behav*. 2011;36(4):400-3.

Records from 629 smokers who had sought help from five selected Stop Smoking Clinics were analysed. Consistent with the findings from other countries, older age and longer duration of previous quit attempts were associated with successful smoking cessation. Greater baseline carbonmonoxide readings, but not Fagerstrom Test for Nicotine Dependence, predicted failure to quit at six-month.

Only half of the referred cases of cutaneous larva migrans had correct diagnosis at referral

Yap FB. Cutaneous larva migrans in Hospital Kuala Lumpur, Malaysia: rate of correct diagnosis made by the referring primary care doctors. *Trans R Soc Trop Med Hyg*. 2011;105(7):405-8.

This is a retrospective study of 31 cases of cutaneous larva migrans seen in the Department of Dermatology, Hospital Kuala Lumpur. Pruritus was reported in 83.9% of cases and serpiginous tracts in 100%. The mean lesion count was 4.4 and the mean duration of disease before presentation was 3.1 weeks. The majority of skin lesions were on the buttock and lower extremities. Only 45.2% of patients had the correct diagnosis made by the referring primary care doctors.