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| TABLE Incidence of anencephaly and other neural tube defects in Jahra Hospital, consecutive births 1985 to 1987. |
|---|---|---|---|---|
| 1985 | 1986 | 1987 | Overall | Per 1000 total births |
| Total births | 7360 | 7033 | 7553 | 21946 |
| Anencephaly | 6 | 11 | 5 | 22 |
| Occipital encephalocele | 1 | — | 2 | 3 |
| Spina bifida cystica | 4 | 4 | 5 | 13 |
| Total NTDs | 11 | 15 | 12 | 38 |
| Total births | 7360 | 7033 | 7553 | 21946 |

programme was encouraging. During the next three years from January 1985 to December 1987, 22 anencephalic babies were ascertained among 21,946 consecutive deliveries in Jahra hospital (table) with a significant decline in the prevalence from 2.05/1000 cases in 1983 to 1/1000 total births (p<0.01).

In the absence of selective therapeutic abortion of malformed babies, on religious grounds, the significant decline in birth prevalence of anencephaly among Bedouin women, in apparent association with the introduction of mass dietary education, offers further evidence of a nutritional contribution to the causation of NTDs. It is difficult to believe that the effect might simply be a ‘natural’ decline in prevalence. It can more likely be attributed to the improved quality of food of the Bedouins and to higher consumption of fresh vegetables and fruit, rich in folic acid, in addition to their traditional foods, rice and meat.

We expect a progressive fall in the prevalence of anencephaly in the Bedouin population with the improvement of the mother’s diet, leaving a baseline prevalence owing to more ‘genetic’ factors, which, especially in a population such as the Bedouins, would well include some autosomal recessive cases.3 8

Talaat I Farag*, Sadika A Al-Awadi*, Salem Yassin†, Talaat A El-Kassaby†, Shaheena Jafary†, R Ushaa†, R Umat†, Samir A Mady†, Mohammed Fahrr†, Mohammed Mannae†, Serikanthie Senartne†, and Tahsin S Khan‡

*Kuwait Medical Genetics Centre; and Departments of Gynaecology and Obstetrics† and Neonatology‡, Jahra Hospital, PO Box 31145, Sulibikhat 90802, Kuwait.

References


Autosomal dominant sneezing disorder provoked by fullness of stomach

SIR,

At a birth defects meeting in 1978, four physicians1 described an autosomal dominant disorder of nearly uncontrollable paroxysms of sneezing provoked in a reflex fashion by the sudden exposure of a dark adapted subject to intensely bright light, usually sunlight. The four physicians were themselves the index subjects of the four families they reported. Subsequently, another instance of autosomal dominant transmission was documented.2 The authors suggested that sneezing in response to light, or photic sneeze reflex, as designated earlier,3 is a common yet poorly understood phenomenon. They found it in nine out of 25 neurologists at Johns Hopkins Hospital, but only two knew that such a specific reflex existed.

We report a documented family history of another sneezing disorder, similarly transmitted in an autosomal dominant manner, but provoked by fullness of the stomach. In the index subject, who is a
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phenotypically normal 32 year old man, fullness of the stomach immediately after meals invariably results in three or four uncontrollable sneezes. This phenomenon is also present in his three brothers, one of his two sisters, his father, an uncle and his son, and the grandfather. The index subject became curious when his daughter started to show the phenomenon at the age of one year, and he mentioned it to one of us when he was examined for an unrelated skin lesion. The 'stomach sneeze reflex' in this family has no relation to the type of food and occurs only when the stomach is full to the extent that no more can be eaten. There are usually three or four sneezes but may be as many as 15 consecutive sneezes.

No-one in this family has persisting otolaryngeal or allergic disease or other conditions known to produce paroxysmal sneezing. 4 5 In addition, photic sneeze reflex was denied completely.

Although there is no evidence of such a phenomenon in medical, physiology, or neurology textbooks, we think it may not be uncommon, but is unrecognised mainly because it causes no annoyance. However, after discussing this phenomenon with 22 colleagues we learnt about a further three sporadic cases. We hope that further studies will clarify how many of us sneeze uncontrollably after heavy meals and why.

AHMAD S TEEBI AND QASEM A AL-SALEH
Kuwait Medical Genetics Centre,
Maternity Hospital, and
Dermatology Unit, Farwania Hospital,
PO Box 36660, Raas 24757, Kuwait.

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