Incidence of Infantile Hypertrophic Pyloric Stenosis in Jordan

Sir,

Infantile hypertrophic pyloric stenosis (IHPS) is reported to be more common in Whites as in the UK and USA than in Blacks,1,2 those of Chinese origin3 and in India.3

We carried out a study to investigate this condition in the north of Jordan and compare it with international figures. From early 1985 till mid 1990, 108200 live births were registered in the north of Jordan, and the male to female ratio was nearly equal. During the same period 26 cases of IHPS were diagnosed and treated surgically after being referred to our hospital—the only referral hospital in the north of Jordan for such cases—thus giving an incidence of approximately 1:4150 live births (0.24:1000 live births).

The average age was 31 days (range 18-60 days) except in one case which was diagnosed 16 h after delivery and operated upon at 30 h after delivery. There were 20 males and six females (M:F 3.3:1); only four cases were first born, while the others ranged from the third to the seventh born, and no definite history could be obtained which suggested a familial pattern in any of the cases.

Sixteen cases occurred in spring (April to June), six cases in autumn (September to November) and four cases in summer (July). The number of cases fluctuated between three and six cases per year.

In Whites IHPS varies from 1 to 8.8:1000 live births.1 The highest incidence was reported from Scotland.4 In Malaysia it is about 1:3500 live births4 and in India6 1:6000 live births. So it seems that the incidence in Jordan lies between those figures from India and Malaysia, but less than those from the White societies. Also our study could not confirm any familial incidence, nor any evidence of the condition being more common in first-born babies.

Due to the small number of cases the male to female ratio and the average age might not be significant. What is shared with other series is that the seasons of the year with the highest incidence are spring and autumn,7 in our study around 85% of the cases occurred in these seasons.

References
5 Joseph TP, Nair RR. Congenital pyloric stenosis (9 years clinical study of 42 cases). Ind J Surg 1974; 36: 221-223.

Temporary Reduction of a Diaphragmatic Hernia by Accidental Pneumothorax in the Neonate

SIR,

Diaphragmatic hernia is present in approximately 1 in 4000 live-born infants and is usually an urgent clinical problem.1 The cardinal clinical features are well described and diagnosis is usually made by a chest X-ray, usually without the aid of contrast medium.2 We recently observed an infant in whom tension pneumothorax reduced the diaphragmatic hernia delaying the diagnosis by several hours. To our knowledge, a similar experience has not been reported previously.

A male infant, weighing 2.31 kg was delivered by Caesarean section for fetal distress at 34 weeks of gestation to a gravida 6, para 5 mother. The mother was hospitalized for 3 weeks before delivery, with ruptured membranes and pyrexia and was receiving treatment with antibiotics.

At birth, the infant failed to breath spontaneously. He required orotracheal intubation and positive pressure ventilation for a few minutes. Subsequently, he needed positive pressure ventilation to maintain satisfactory blood gases. After initial improvement he suddenly deteriorated while on the ventilator with loss of colour and oxygen saturation. A chest radiograph showed a left-sided tension pneumothorax. Ventilator malfunction was the cause of the tension pneumothorax, as the pressures were reading high (PEAK 60 cm, PEEP 10 cm of water). The pneumothorax was drained with improvement in the baby's condition. A repeat radiograph 2 h later showed the abdominal contents herniated into the left hemithorax establishing the diagnosis of a left diaphragmatic hernia. At surgery, a large defect in the postero-lateral part of the diaphragm noted (foramen of Bochdalek). The stomach, some of the intestines and the spleen had herniated through it. The infant underwent a successful repair of the diaphragmatic hernia.

*Correspondence
Infants with diaphragmatic hernias fall into three distinct groups of pulmonary hypoplasia: minimum, unilateral and bilateral. The pneumothorax in diaphragmatic hernia results from high ventilatory pressures coupled with pulmonary hypoplasia. The pneumothorax in our case was clearly because of the malfunction of a mechanical ventilator. The high pressure (60 cm of water) reduced the herniated contents from the chest. In the presence of a hernial sac the pneumoperitoneum membranes usually do not develop. The abdominal contents reherniated when the pressure in the chest was reduced by drainage of the pneumothorax. Charles H. Mayo in 1927 at a meeting of the American Surgical Association, while relating the treatment of diaphragmatic hernia suggested that deliberate creation of pneumothorax facilitates reduction of herniated contents. Although pneumothorax is known to occur with hypoplastic lungs with positive pressure ventilation, reduction of diaphragmatic hernia because of pneumothorax seems to be unique to this patient.

**SUDHAKAR BARARAO PATIL MBBS MRCP DCH Consultant Paediatrician, Department of Paediatrics, King Faisal Military Hospital, PO Box 101, Khamis Mushayt, Saudi Arabia**

**ANIL KUMAR GARG MBBS MRCP Consultant Paediatrician**

**Saudi Medical Journal Volume 13 No. 1 January 1992:** 78

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### Table 1

<table>
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<tr>
<th>Tribe</th>
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### References


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**α-Thalassaemia, Blood Groups and Saudi Tribes**

SIR,

The article by Dr Niazi et al.\(^1\) is a useful contribution to our knowledge of red cell abnormalities in newborns of Saudi Arabian National Guard dependents but I am concerned that they have used Hb Barts to estimate the proportion of the population who have α-thalassaemia. Studies which depend on the detection of Hb Barts in cord blood have been shown to underestimate the frequency of α-thalassaemia.\(^2\) My colleagues and I found Hb Barts in 4% of cord bloods from another group of Saudi National Guard dependents.\(^3\) However, DNA studies\(^4\) on the same newborn population showed that the frequency of the single α-gene defect (-α) form of α-thalassaemia was 0.13 so that the calculated total with the defect (heterozygotes plus homozygotes) is 24%. This demonstrates the importance of using DNA studies, rather than Hb Barts, to obtain accurate estimates of the frequency of α-thalassaemia.

Niazi et al. wisely avoided making any generalizations about the incidence of various red cell abnormalities in the Saudi population and correctly drew attention to variations in different parts of the Kingdom. DNA studies\(^5\)\(^-\)\(^9\) have shown marked variation in the frequency of α-thalassaemia in different parts of Saudi Arabia and this is probably due to differences in the historical exposure to malaria. El-Hazmi\(^10\) carried out an interesting study in which he found differences in genetic red cell abnormalities between villages in quite close proximity. If separate tribes were examined using DNA techniques, differences in the frequency of α-thalassaemia would be likely which might reflect the historical prevalence of malaria in their original homelands. The population which we studied were all from Saudi Bedouin tribes living in the area around Jeddah but whose original homelands were from a wide area of Saudi Arabia. Your readers may be interested in the gene frequency of A, B, O and Rhesus (D) in some of these tribes (Table 1). The results suggest that there are some genetic differences between tribes and illustrate the difficulty of making generalizations about the frequency of genetic abnormalities in the Saudi population.

**DAVID STEVENS FRCP**

**Pediatrian Department, Gloucestershire Royal Hospital, Gloucester, UK,**

**Saudi Medical Journal February 1992:** 78

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### References


SIR,

I have read with interest the above short communication by Dr Stevens describing the prevalence of α-thalassaemia and gene frequency of A, B, O and Rh (D) in the various tribes of Saudi Arabia serving the National Guard force. It is interesting to note that the incidence of α-thalassaemia in our newborns (5%) was almost (4%) as observed by screening in another group of Saudi Arabian National Guard dependents. I fully agree with the statement that several studies including a very large study from the eastern province of Saudi Arabia on cord blood screening for erythrocytic genetic disorders\(^1\) have shown
that using elevated levels of Hb Barts > 2% as a marker for α-thalassaemia grossly underestimates the frequency. The reason behind this discrepancy is the use of inappropriate methodology. I have pointed out on several occasions that the cellulose acetate haemoglobin electrophoresis commonly available in most hospital laboratories is unsatisfactory in detecting the levels of Hb Barts less than 2% and for this reason more sophisticated techniques which are not difficult to establish are required. These include isoelectric focusing (IEF), high performance liquid chromatography (HPLC) and radioimmunoassay (RIA). Out of these, RIA is most sensitive and can accurately detect as little as 0.05% of Hb Barts. This has been clearly demonstrated in a study from the Medical College of Georgia. I have personal experience with RIA and it is extremely accurate and sensitive for the identification and quantitation of several normal and abnormal haemoglobins and can be used for mass screening. In the case of Hb Barts, the antibodies for this abnormal haemoglobin can be made in New Zealand white rabbits and the sera (anti-Hb Barts) can be rendered specific for γ tetramer by immunaoabsorption with Hbf-Sepharose conjugate. Because of sensitivity and accuracy, RIA can resolve the issue of the prevalence of α-thalassaemia in a given population, and DNA analysis which is still a research tool can be performed to show what type of α-thalassaemia is present. I am optimistic that by simultaneously estimating the levels of Hb Barts by RIA and subjecting same specimens for DNA analysis we can derive some relationship between the levels of Hb Barts and the number of α-globin genes deleted. Data are available to show that Hb Barts levels > 0.6% indicate the presence of all α-thalassaemia gene. Dr Stevens has rightly pointed out the differences in the prevalence of α-thalassaemia detected by two methods in his study (4% by routine electrophoresis vs 24% by DNA analysis). We had a similar experience in our study from eastern Saudi Arabia. We estimated that 30–40% of newborns in the Qateef area had Hb Barts > 2% and the subsequent DNA analysis showed that the actual incidence of α-thalassaemia was around 60%. We are now performing DNA analysis on our Saudi Arabian National Guard infants and preliminary data indicate that the reported incidence of 5% α-thalassaemia in our newborns is an underestimation. These data will be published when the study is complete.

GULZAR AHMED NIAZI PhD
Haematologist,
Department of Medical Education and Research,
King Fahad National Guard Hospital,
PO Box 22490, Riyadh 11421, Saudi Arabia

References

Neonatal Hydrocolpos

Sir,

Neonatal hydrocolpos secondary to vaginal obstruction is an uncommon condition and is frequently misdiagnosed due to lack of awareness of this condition. We present a report to highlight this.

A full-term female Saudi baby was referred for abdominal distension. Preliminary clinical and ultrasound evaluation at the referring hospital had suggested retention of urine. On examination the baby was healthy and active. There was an abdominal mass extending from the epigastric region to the pelvis. The superior and lateral borders were well defined but it was not possible to get below the mass. The mass was firm, cystic and laterally mobile. On spreading the vulva a glistening whitish membrane was seen to bulge from the vagina making the diagnosis obvious. An ultrasound examination showed a large fluid-filled swelling arising from the pelvis. A cystogram showed the bladder was compressed anteriorly. The baby was treated by a simple cruciate division of the fleshy hymen.

Neonatal hydrocolpos is secondary to distal vaginal obstruction or stenosis. It can occur as a single anomaly or with other anomalies such as McKusick–Kaufman syndrome or as part of a cloacal anomaly.

The vaginal obstruction can be a simple imperforate hymen or vaginal atresia which may be high or low. A high index of suspicion and detailed genital examination will immediately prove the diagnosis in cases of imperforate hymen which occurs more commonly.

Ultrasound is a useful tool but is frequently misleading as the bladder is compressed anteriorly and not visualized, leading to a mistaken diagnosis of the bladder distention. This can be easily overcome by filling the bladder with contrast and taking lateral X-rays.

The management depends on the level of obstruction. It is stressed that a regular follow-up is needed to assess the vaginal opening as well as any collection in the vagina due to reflux of urine.

GHULAM NABI MD
Paediatrician,
Department of Paediatrics,
Khamis Civil Hospital,
PO Box 737,
Khamis Mushayt

JAYANTH KRISHNA IVENGAR MS MCH
Consulting Paediatric Surgeon,
Asir Central Hospital,
ABHA


References
Serum Zinc Levels in Saudi Patients with Chronic Liver Disease

Sir,

I have read the two letters on serum zinc levels in Saudi patients with chronic liver disease but find no mention of the relationship between serum zinc and serum albumin. The observations made in your journal may be less confusing to the authors if this factor is considered. I suggest that they refer to the paper on relation between zinc status and hepatic functional reserve in patients with liver disease by Goode, Kelleher and Walker (Gut 1990; 31: 694–697) in which the question of zinc depletion in patients with liver disease is examined.

MARGARET E. ELMES MBBS FAD
Department of Pathology,
Heath Park
Cardiff CF4 4XN,
Wales, UK