Is the incidence of gastroschisis rising in South Africa in accordance with international trends?

A retrospective analysis at Pretoria Academic and Kalafong hospitals, 1981 - 2001

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Summary

Objectives. Analysis of the incidence of gastroschisis compared with the incidence of exomphalos as a percentage of total paediatric surgical admissions.

Design. Retrospective observational analysis using data from the ward admissions registers of the paediatric surgical wards of Pretoria Academic and Kalafong hospitals and from the weekly statistics sheets and audits thereof of the Paediatric Surgery Department at Pretoria Academic Hospital. Patient files from Pretoria Academic were used where available to confirm data.

Setting. Pretoria Academic Hospital (PAH) and Kalafong Hospital (KH), Pretoria, from March 1981 to December 2001, excluding Kalafong Hospital data from February 1984 to 12 June 1984 and for 2001.

Results. Forty-eight cases of gastroschisis and 139 cases of exomphalos were seen at PAH and KH out of 21 495 total paediatric surgery ward admissions. The average incidence of gastroschisis increased 35-fold from the 7-year period 1981 - 1987 to the 7-year period 1995 - 2001, while the average incidence of exomphalos compared across the same periods only showed a 1.82-fold increase. The incidence of gastroschisis rose above that of exomphalos in 1999 and remained so through to 2001. The incidence of exomphalos showed a general rising trend from 1981 to 2001 except for a sudden unexplained increase in 1995 and 1996.

Gastroschisis presenting at PAH was associated with a mortality rate of at least 38.7%. Reasons for this high mortality rate cannot be adequately evaluated owing to poor record keeping.

Conclusions. The incidence of gastroschisis presenting at PAH and KH has risen dramatically relative to the incidence of the macroscopically similar exomphalos over the period 1981 - 2001, with the incidence of gastroschisis rising above the incidence of exomphalos. A larger scale study looking at birth prevalence of gastroschisis and exomphalos in South Africa is necessary to determine whether this represents a true differential change in the incidence of gastroschisis, reflecting internationally observed increases in the birth prevalence of this defect, or whether it is merely due to logistical changes within the administration of the Department of Health serving the area.

Background

The overall worldwide birth prevalence of gastroschisis has risen from 0.29 per 10 000 births in 1974 to 1.66 per 10 000 births in 1998.[1] This increase has been observed since the 1970s but began at the end of the 1980s in several areas.[1-5] There is a wide regional variation in the birth prevalence of gastroschisis, with the incidence in Japan being 0.467 per 10 000 births for 1996 - 1997,[2] compared with 4.4 per 10 000 births in South East England in 1995.[3] The incidence of the macroscopically similar exomphalos (omphalocele) however, has remained stable or decreased.[4]

The causation of gastroschisis remains uncertain, although the interruption of the fetal omphalomesenteric arterial supply has been suggested as the pathophysiology.[6] Teenage mothers have been shown to be at significantly higher risk of having a child with gastroschisis, suggesting a lifestyle exposure to an environmental teratogen. Maternal use of recreational drugs, including cocaine, marijuana, amphetamines and alcohol, as well as maternal smoking, increases the risk of gastroschisis.[4] However, no significant changes in the rates of maternal drug use, tobacco smoking or alcohol abuse, or of conception while taking the oral contraceptive pill, have been found to correlate with the sudden and rapid increase in the incidence of gastroschisis.[3]

The causation of gastroschisis is probably multifactorial.[4] The sporadic incidence and low familial recurrence rate make a genetic cause unlikely[7] although an interaction between a specific teratogen and an environmental or genetic predisposition has been suggested. The rapid rate of increase in incidence and the wide regional variation suggest an environmental or nutritional cause.[1,3] A teratogen with a high exposure rate and low efficacy is likely, as no unusual potential teratogens have been identified in case history or case control studies.[7] A link to influenza infection during the first trimester of pregnancy, with an unusually high rate of children born with gastroschisis in the first quarter of the year in England, has been proposed.[3]

Method

Data from the ward admissions registers of the paediatric surgical wards of Pretoria Academic Hospital (PAH) and Kalafong Hospital (KH), and from the discharge records and the weekly statistics sheets and audits thereof of the Paediatric Surgery Department at PAH, were collected and analysed. Patient files from PAH were used where available to confirm

data. The 21-year period from 1981 to 2001 was divided into three 7-year periods, and the average incidence of gastroschisis and exomphalos (as a proportion of total paediatric surgery admissions) for each 7-year period was determined for comparison. Data for KH are missing for a 4-month period during 1984 and for 2001. No data were available for the first 3 months of 1981 for either hospitals. The referral area served by the hospital is ill-defined and has changed over the past 2 decades owing to changes in the regional health infrastructure, so it was impossible to ascertain the total number of births in the large region out of which the cases of gastroschisis and exomphalos were seen.

Data on repair of gastroschisis at PAH (primary repair or silo closure with secondary repair) and survival were collected, but inadequate record-keeping means that this information is very much incomplete. Records of ward admissions were made by ward clerks and nursing sisters who are often insufficiently trained and are incomplete and often inaccurate. Cross-checking between registers, patient files and statistics sheets kept by the doctors was done as much as possible.

Results and discussion

Out of the total of 21 495 admissions to the paediatric surgery wards for the period 1981 - 2001, 48 cases of gastroschisis and 139 cases of exomphalos were found.

The number of cases of exomphalos showed a general rising trend (with an average increase of a factor of 1.23 cases a year) from 1981 to 2001. However, there was a sudden peak in the number of cases of exomphalos seen at KH in 1995 and 1996, with 27 cases out of 1 499 paediatric surgical admissions (1.8%) recorded, compared with 54 cases out of 10 835 admissions (0.5%) over the preceding 14 years. No similar peak in the incidence of exomphalos at PAH was seen and the number of cases of abdominal wall defects at Ga-Rankuwa Hospital (now Dr George Mukhari Hospital), the nearest other state hospital with a paediatric surgical unit, was remarkably constant over this period. No explanation for the peak in cases of exomphalos was found. It could represent improved transport or perinatal care in the local surroundings or it could be an incidental finding. In the light of the overall rising trend in exomphalos it does not appear significant.

The average incidence of gastroschisis showed a 7-fold increase from the first 7-year period (1981 - 1987 — average incidence 0.02%) to the second 7-year period (1988 - 1994 — average incidence 0.11%), and a further 5-fold increase in the third 7-year period (1995 - 2001— average incidence 0.55%). In comparison, the average incidence of exomphalos increased by 1.3 from the first 7-year period (average incidence 0.47%) to the second (average incidence 0.62%) and a further 1.4-fold (to 0.89%) during the third 7-year period.

The average incidence of gastroschisis therefore increased 35-fold from the first to the third 7-year period, compared with a 1.82-fold increase in the average incidence of the macroscopically similar exomphalos over the same period. The incidence of gastroschisis rose and remained above that of exomphalos in 1999 through to 2001 (Fig. 1).

This finding could reflect a real increase in the incidence of gastroschisis, as the exogenous factors affecting the presentation of a birth defect like gastroschisis would be presumed to affect exomphalos to the same degree. Of the total 48 cases of gastroschisis, 32 (67%) were seen at PAH and KH over the 3-year period 1999 - 2001. (This excludes 2001 data for KH, which were not available.) The increasing trend in exomphalos over 1999 - 2001 remained consistent with the overall rising trend-line of exomphalos.

No demographic data on maternal age were available. This is unfortunate, as a comparison with the international inverse age-related risk profile could be of aetiological significance.[1-4]
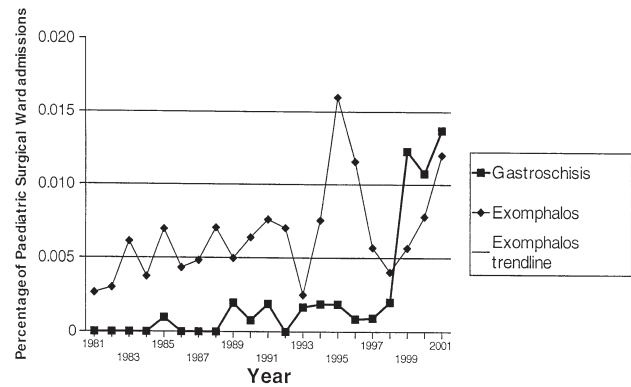

Fig. 1. Incidence of gastroschisis compared with exomphalos, Pretoria Academic and Kalafong hospitals, 1981 - 2001.

Survival rate of gastroschisis cases at PAH

Thirty-one new cases of gastroschisis were seen at PAH from March 1981 to December 2001. Figures for survival are incomplete but show that at least 12 (39%) patients with gastroschisis out of the 31 seen died. Mortality in the First World is under 10% for gastroschisis thanks to antenatal diagnosis, surgical intervention soon after birth and availability of neonatal intensive care units (NICUs) and total parenteral nutrition (TPN).[8-10] A study in Nigeria, where these are not available and late presentation is the rule, reported a 71% mortality,[11] while mortality at Red Cross War Memorial Children's Hospital in Cape Town from 1970 to 1977 was 31%.[12]

Data on reasons for mortality were not available. However, intensive care and TPN is limited at PAH because of lack of paediatric ICU beds and budgetary constraints. The high mortality rate also reflects the poor quality of the regional perinatal care and referral system, with many cases presenting to tertiary care late. Complications such as hypothermia, acidosis, dehydration and bowel oedema that result from this time delay can increase mortality.

Information on the surgical procedure used to close the abdominal wall defect were incomplete, with the procedure used being unknown in 11 out of 31 cases (35%). However, a Gortex or silastic pouch was used with secondary closure of the defect in at least 11 cases (35%). These figures (albeit incomplete) show a slightly higher rate of secondary closure compared with Indiana, USA, where 31% of cases were closed secondarily.[8] At Red Cross Children's Hospital from 1960 to 1977, reinforced silastic (Gortex) closure was used in 40% (with a 40% mortality), while 56% were treated with primary repair (with a 50% mortality), and in the remaining cases skin closure only was done.[12]

Co-morbidity as a factor contributing to mortality was impossible to evaluate. No records of gestational age were kept and data on birth mass were only available for a few infants. Two cases of concomitant small-bowel atresia, but no record of other concomitant major anomalies, were found. It was noted that one patient who died was HIV-positive, but notes on the case were inadequate to link this to the cause of death.

Conclusion

The incidence of gastroschisis at both PAH and KH has significantly increased over the 21-year period 1981 - 2001, to above the incidence of exomphalos, while the macroscopically

similar exomphalos, used as a control for exogenous factors such as regional health referral and transport infrastructure changes, has shown a relatively constant rate of increase over the same period except for a sudden peak in 1995 and 1996. Socio-economic and political factors have changed and affected the health infrastructure in South Africa significantly over the past decade. It is therefore difficult to assess whether there has been a real increase in the incidence of gastroschisis, whether it is merely a sporadic and incidental finding such as the peak in exomphalos seen at KH in 1995 and 1996, or whether it is due to improved referral services in the hospital feeder areas. Further study looking at the wider South African picture and examining the demographics of abdominal wall defects, including maternal age distribution and possible aetiological factors, is necessary. Better epidemiological record-keeping, including an efficient national registry for birth defects, would significantly aid research on congenital abnormalities in South Africa and thus improve their management.

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