Wiedemann–Beckwith syndrome in one of monozygotic twins

SIR,

The article by Bose and his colleagues1 deserves some comment. It is certainly unusual to find an exomphalos in one of monozygotic twins, although it has been described before. To label a baby as Beckwith–Wiedemann syndrome simply on the basis of the presence of an exomphalos and a large tongue is surely, however, not justified. The essence of the syndrome is the combination of exomphalos, macroglossia, and gigantism, manifest as both somatic and visceromegaly, together with the sporadic occurrence of other features such as indented ear lobes and naevus flammeus. The definitive histological diagnosis is based on the finding of cytomegaly of the adrenal cortex. In the absence of some corroborative findings it would be more reasonable to consider this baby as suffering from exomphalos, complicated by the incidental finding of macroglossia.

I am also interested in the authors’ use of the term Wiedemann–Beckwith syndrome. Since the syndrome was described by Beckwith in 1963 (Beckwith JB. Extreme cytomegaly of the adrenal fetal cortex, omphalocele, hyperplasia of the kidneys and pancreas, and Leydig cell hyperplasia—another syndrome? Presented at annual meeting of Western Society for Pediatric Research, Los Angeles, California, 1963.) and by Wiedemann in 1964,2 then for both chronological and alphabetical reasons it would seem reasonable to use the well established and recognised terminology of Beckwith–Wiedemann syndrome. Perhaps, however, the authors can explain the justification for this change.

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Drs Bose and Forsyth comment:

We thank Mr Brown for his interest in our case report.1 Although exomphalos, macroglossia, and gigantism are common features of Wiedemann–Beckwith syndrome they are by no means constant. In the review of 49 cases of the syndrome by Fillipi and McKusick, 4 which included the original cases reported by Wiedemann2 and Beckwith,4 macroglossia was present in 95% of cases, exomphalos in 93%, hypoglycaemia in 82%, visceromegaly in 79%, and macrosomia in 74%. Birth weights of 12 infants in this series,3 born at term, ranged from 2900 g to 5675 g, with a mean value of 4050 g. Comparable figures were found by Irving5 with a mean value of 3857 g as her series included preterm infants as well. The infant that we described1 had macroglossia that eventually required partial glossectomy, exomphalos, hypoglycaemia, and, if compared with her identical twin, has appreciable macrosomia, which is still present at the age of 2 years. We therefore feel that there is sufficient evidence to support our clinical diagnosis of Wiedemann–Beckwith syndrome.

Provided that both investigators2, 4 are acknowledged we do not have strong views on the terminology used to label the syndrome. We note that Beckwith first described autopsy findings of this syndrome at the annual meeting of Western Society for Pediatric Research in 1963, and subsequently in two living children in 1964 at the same time as Wiedemann’s formal publication.2

References


Transcutaneous oxygen and carbon dioxide monitoring in intensive care

SIR,

Determination of end tidal carbon dioxide in children who require respiratory support is non-invasive, more closely related to arterial carbon dioxide, and more convenient than determination of transcutaneous carbon dioxide tension.

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United States

Dr Helms comments:

Dr Singh suggests that end tidal carbon dioxide is less invasive and more closely correlated to arterial carbon dioxide than transcutaneous measurements. This has not been our experience, as the accompanying Figure shows, particularly in children with lung disease. The relation of end tidal carbon dioxide to arterial carbon dioxide depends on the evenness of gas mixing within the lung and the time available to reach a true end tidal plateau. Our data suggest that this is rarely the case in children with lung disease. We have found the end tidal carbon dioxide technique very useful, however, in the management of children with raised intracranial pressure and with normal lungs. It is in this latter group that episodes of intracranial...
hypertension can be brought under control by hyper-ventilation.

Figure  Relation between end tidal carbon dioxide and arterial carbon dioxide in 17 children with lung disease (open circles) and 42 children with radiologically normal lungs (closed circles). The line of identity is shown. Fifteen observations hidden. Data courtesy of D P Heaf.

Synchronising breathing and positive pressure ventilation

Sir,

The paper by Field et al1 shows what we have observed during artificial ventilatory support: shorter inspiratory times and faster respiratory rates result in more synchronised breathing, decreasing the need for sedation or paralysis.

Greenough et al2 incriminated asynchronous breathing in the genesis of pneumothorax and referred to ‘active expiration against the ventilator’ as being the main factor. We would suggest that ‘deep inspiration with the ventilator’ is more likely to be the problem. It is at this time in the ventilator cycle that the additive effect of ventilator pressure and negative pleural pressure results in a large transpulmonary pressure and maximal alveolar distension. Active expiration against the ventilator results in a smaller transpulmonary pressure and less alveolar distension: an unlikely time for alveolar rupture. Both of these effects can be seen in Fig. 1 of Field’s paper.

In addition to its disruptive effect on ventilation and its relation to pneumothoraces, breathing out of synchrony also results in the type of fluctuations in the cerebral and systemic circulations that have been associated with an increased risk of intraventricular haemorrhage by Perlman et al.3

Synchronised breathing achieved by higher rates and shorter inspiratory times would also have the advantage over paralysis of better distribution of ventilation through the continued action of the diaphragm.

References

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Immunoregulatory treatment for minimal change nephrotic syndrome

Sir,

It is likely that the mechanism of action of levamisole in the treatment of minimal change nephrotic syndrome as reported by Mehta1 and Tanphaichitr2 is specific suppression of excessive suppressor T cell function.

All drugs used in the treatment of minimal change nephrotic syndrome, including glucocorticoids, cyclophosphamide, nitrogen mustard, and azathioprine, have profound immunosuppressive actions except levamisole, which as shown in the Mehta study and in others is an immunostimulant. There are two paradoxes that need explanation:

(a) The treatment of a disease in which immunosuppression is an intrinsic part with drugs that suppress the immune system further.

(b) The successful treatment of minimal change disease with several immunosuppressive drugs and one immunostimulant drug.

The answers to these paradoxes can be found by more detailed study of the cellular actions of levamisole. Levamisole achieves its immunostimulant effects by specifically suppressing suppressor T lymphocytes.3 4 The other drugs suppress T lymphocytes non-specifically along with other lymphocyte subgroups.

Excessive suppressor T lymphocyte function in untreated minimal change nephrotic syndrome might cause both the disease through release of a lymphocine, which disrupts anionic binding sites, and the observed depression of immune function. Further investigation of this interesting treatment is most certainly appropriate, both to give insight into the disease and to develop a more specific form of treatment.

References
1 Mehta KP, Ali U, Kutty M, Kolhatkar U. Immunoregulatory
Dr Helms comments

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Updated information and services can be found at:
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