

Letter to the editor

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Tumours and hemihypertrophy associated with Wiedemann-Beckwith syndrome

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Sir, — The increased danger of tumours developing in children with the exomphalos-macroglossia-gigantism syndrome (Wiedemann-Beckwith syndrome) is well known. An analysis of cases in the literature and our own observations made up to 31 July 1983 yielded the following results. Of 388 children, 29 developed a total of 32 neoplasms (3 double tumours or second tumours). Of

these tumours, 26 were localized intra-abdominally, 14 being Wilms tumours and 5 adrenal carcinomas (Table 1). Therefore, the tumour rate is 7.5% (mainly malignant). A hemihypertrophy, either partial or complete, was noted in 12.5% of the total number of children, but in >40% of the children with neoplasms. Every child with this syndrome needs to be examined carefully with renal sono-

Table 1

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|----------------------------|----------------------------|
| <i>Intra-abdominally:</i> | 14 Nephroblastoma |
| | 5 Adrenocortical carcinoma |
| | 2 Hepatoblastoma |
| | 5 Other |
| <i>Extra-abdominally:</i> | 2 Intracranial malignoma |
| | 3 Other |
| <i>Malignant lymphoma:</i> | 1 |

graphy, first at 3-month intervals and after the 3rd year of life at 6-month intervals.

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