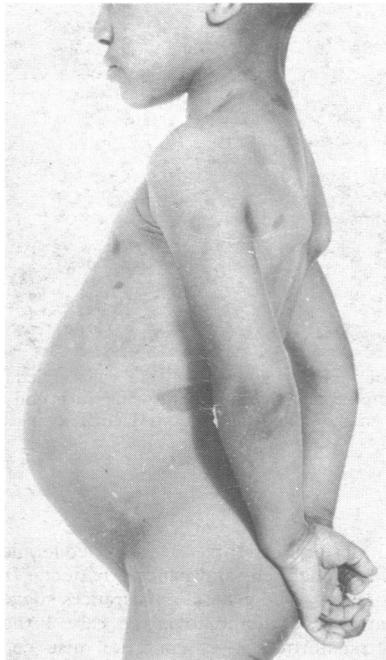


Wilms's tumour and neurofibromatosis

The association between Wilms's tumour and certain congenital anomalies is well documented. An increased incidence of aniridia, hemihypertrophy, and abnormalities of the genitourinary tract has been described.¹ We report the case of a child with Wilms's tumour and neurofibromatosis.

Case report

A 3-year-old Negro girl was admitted on 10 June 1975 with a six-month history of a swelling in the abdomen. Physical examination showed widespread café-au-lait spots over the chest, trunk, and arms (see figure) and a swelling on the wrist thought to be a neurofibroma. A large smooth mass



The patient, showing widespread café-au-lait spots.

was palpable in the left hypochondrium. The patient's father, a paternal aunt, and the paternal grandfather all had neurofibromatosis. An intravenous pyelogram showed distortion of the left kidney with displacement by a soft mass extending across the mid-line. The chest x-ray film and a skeletal survey were normal. Vanillylmandelic (VMA) acid excretion was within normal limits. On 20 June, a large nephroblastoma (15.5 × 10.5 cm), together with the left kidney and 6.5 cm of the ureter, were removed; to the naked eye the tumour had not extended beyond the capsule of the kidney. Between July 1975 and July 1976 she received 11 courses of chemotherapy with Actinomycin D and vincristine. She remains in remission with no evidence of recurrence.

Discussion

Wilms's tumour is the second commonest intra-abdominal neoplasm in childhood: the incidence is roughly one case per 200 000 children per year.² It is more common in patients with certain developmental anomalies, occurring in about a third of patients with congenital aniridia.³ Ipsilateral or contralateral hemihypertrophy is also commonly associated with this tumour. Other urinary tract abnormalities have been described in Wilms's tumour, including hypospadias, cryptorchidism, horseshoe or multicystic renal disease, duplicated ureters, renal hypoplasia, and pseudohermaphroditism.

Neurofibromatosis occurs in 1 per 3000 live births and is characterised by café-au-lait spots and multiple neurofibromas. Inherited as an autosomal dominant characteristic of variable expressivity, it forms one of the group of hamartomatous disorders in which there is

localised excess of growth and occasionally neoplastic change. There is an increased incidence of gliomas, acoustic neuromas, meningiomas, and pheochromocytomas. Familial aggregations of tumours of neurogenic origin have been described but only a few reports cite an association between Wilms's tumour and neurofibromatosis.^{1, 4} Although the incidence of congenital anomalies is much greater in bilateral cases of Wilms's tumour,³ the occurrence of two neoplasms in which a genetic influence is particularly strong is probably more than coincidental. Familial cases of Wilms's tumour have been described without expression in the parent and this has been ascribed to a delayed mutation, possibly viral-induced.⁵

The importance of excluding an underlying Wilms's tumour in any child who presents with one or more of these distinctive congenital anomalies deserves wider recognition in view of the fact that between 80-90% of early cases are now curable.

Requests for reprints should be sent to KDB.

¹ Miller, R W, *Pediatric Research*, 1969, 3, 389.

² Mott, M G, *British Journal of Hospital Medicine*, 1975, 2, 161.

³ Bond, J V, *Lancet*, 1975, 2, 482.

⁴ Kung, F H, and Nyman, W L, in *Cancer Medicine*, ed J Holland, and E Frei III, p 188. Philadelphia, Lee and Febiger, 1973.

⁵ Knudson, A G, and Strong, L C, *Journal of the National Cancer Institute*, 1972, 48, 313.

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Body weight correlates with REM sleep

The fact that REM (rapid eye movement) (paradoxical) sleep is a time when dreaming occurs¹ has led to a neglect of somatic functions at this time and to unsuccessful attempts to relate an individual's personality traits to his characteristic amount of REM sleep. To obtain a reliable measure of the latter it is important to allow adaptation to the sleep laboratory, to record a large number of nights, preferably consecutive, and to allow adequate time in bed each night.

Methods and results

Sixteen healthy volunteers aged 52-67 (mean 59 years), six men and 10 women, slept in the laboratory every four weeks during a 16-week period and always did so for six consecutive nights, of which the first was for adaptation. Lights-out was from 2215 to 0700 h. At the start and end of each week of attendance they were weighed in light clothing. None of the volunteers were receiving drugs affecting the nervous system and they were asked to abstain from alcohol throughout. The 320 electrophysiological records were scored in terms of the usual international criteria. The amounts of each sleep stage on each night were calculated and also, for each subject, the means for his or her 20 nights. The means of the eight weight measures were also calculated.

It was found that body weight and the percentage of total sleep that was spent in REM sleep were related. Spearman's rank correlation test was employed and $r_s = 0.774$, $t = 4.57$, $P < 0.001$, 2-tailed. Log body weight was also correlated with mean total number of minutes of REM sleep (with a product-moment correlation, $r = 0.643$, $t = 3.07$, $p < 0.01$, 2-tailed). The figure illustrates the simple correlation between body weight and percentage of REM sleep. No correlations were found between body weight and total sleep, slow wave sleep or other sleep variables.

Discussion

When patients with anorexia nervosa gained weight the most significant change in their sleep lay in an increase of REM sleep,² while in a study of acute starvation REM sleep fell significantly.³