THERAPY UPDATE

Childhood neuroblastoma

THE DISORDER

A malignant tumour of embryonal cells in the neural crest; arise in the medulla of the adrenal gland or anywhere along the sympathetic nervous chain from the neck to the pelvis

INCIDENCE

- Third most common tumour among children after leukaemia and brain cancers.
- Cumulative death rate of 70 deaths per million children up to 15 years of age in UK 1986–1990.[1]
- Mortality peaks at 2 years of age.

NATURAL HISTORY

- Children presenting with localised, resectable (stage 1) tumours have a 95% five year survival rate.[2]
- Children presenting with advanced metastatic (stage 3 or 4) disease have only a 25% five year survival rate.[3]
- Tumours occasionally regress spontaneously.[4]

SCREENING TESTS

- Blotting wet nappies in children at or under 1 year of age with filter paper and sending the filter paper to laboratories to analyse the amount of homovanillic acid (HVA) and vanillylmandelic acid (VMA) excreted.
- Subsequent tests: repeat urine test

DIAGNOSTIC TEST

Physical examination, chest and abdominal radiography, and abdominal ultrasound or computerised tomography scan.

TREATMENT

Surgery, chemotherapy, radiotherapy, or high dose chemotherapy with bone marrow rescue.

SCREENING PERFORMANCE

- No randomised controlled trial performed.
- Two large trials in Quebec (Canada) and Germany comparing screened populations with concurrent control populations have been completed. The trial in Quebec screened half a million children at 3 weeks and six months of age and followed them up for 8 years.[5 6] The trial in Germany screened two and a half million children at one year of age with a median duration of follow up of 7 years.[7]

INTERPRETATION OF RESULTS

There is no indication that screening reduces mortality (RR=1.3 (0.9–1.9)) (see table). The twofold increased risk of detecting a tumour indicates that screening is detecting more tumours than would otherwise be identified, causing uneccessary risk and distress to the individuals concerned.

OVERALL ASSESSMENT

Screening is not worthwhile and potentially causes harm.

REFERENCES

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4 **Yoneda A**, Oue T, Imura K, *et al*. Observation of untreated patients with neuroblastoma detected by mass screening: a "wait and see" pilot study. *Med & Ped Onc* 2001;**36**:160–2.
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	Quebec study		German study	Both
Age at screening	3 weeks	6 months	1 year	
Positive rate after two urine tests (per 100000)	9	13	120	
Odds of being affected given a positive result	1:1.2	1:0.73	1:12	
Relative risk of mortality in screened *v* unscreened populations	1.4 (95% CI 0.85–2.3; p=0.10)		1.2 (95% CI 0.7–2.0; p=0.56)	1.3 (95% CI 0.9–1.9); p=0.17
Relative risk of a tumour detected in screened *v* unscreened populations	2.2 (95% CI 1.8–2.6; p<0.001)		2.0 (95% CI 1.6–2.4; p<0.001)	2.1 (95% CI 1.8–2.4; p=0.001)