A new clinical sign in Williams syndrome

Editor,—Williams syndrome is a well recognised condition with typical facies, supra- vular aortic stenosis, mental retardation, and a characteristic personality. 1 In a large series (n=235) 96% of patients demonstrated a deletion of the elastin gene from the long arm of chromosome 7 2 Strabismus is common in Williams syndrome 3 and this may contribute to subnormal binocular vision and reduced stereopsis. In a recent study of 28 patients with typical features and deletion of the elastin gene an interesting sign was noted. On further inquiry it was found to have been present in 30% (n=9) of cases. The observation is that as children they have a great reluctance in changing the surface on which they are walking or playing. A typical example would be going from a tiled to a carpeted surface. The child would stop at the threshold and refuse to proceed. They may then feel out the new surface with either a probing finger or in some cases descend to all fours to confirm the suitability of the next surface. The process of transfer may take several minutes. Patients describe this observation in both indoor and outdoor settings. It would seem that there is a problem in determining depth perception when there is either a new pattern or colour to the surface. A reluctance to proceed may reflect a fear of falling to the next surface. Similar difficulties are experienced in attempts to descend stairs. Another interesting observation in this group is the lack of curiosity on the part of the child in handling new or unexpected surfaces. This is most likely an extension of the previous observation that in this surface is undulating, has a variable visual pattern, and combining these factors that contribute to the uncertainty of the surface. Several of the children experienced great distress when faced with this circumstance. As the children grow older the problem diminishes and most have few concerns in changing surfaces by 8 years of age. This clinical sign has not been described previously in this group and I am unaware of any other paediatric group who demonstrate a similar sign. I would be most pleased to hear from any other groups who may have made similar observations.

Expulsion of ventriculoperitoneal shunt tubing

Editor,—In reply to the letter by Dr Swann on the supposedly unique occurrence of expulsion of ventriculoperitoneal shunt tubing, 1 I would like to describe another case, not of expulsion but extrusion of ventriculoperitoneal tube per rectum. A first twin born by caesarean section on our delivery unit at 26 weeks gestation had a stenosed neonatal course due to a large haemorrhagic hydrocephalus. He subsequently needed a ventriculoperitoneal shunt. He was readmitted at the chronological age of 5 months with a large systolic and diastolic swishing sound over the shunt site. He was suspected of having a shunt infection and was treated with intravenous cefotaxime and fluocoxacin. After 24 hours in hospital the nursing staff noticed, while changing a nappy, extrusion of the shunt per rectum. He was immediately transferred to the neighbouring neurosurgical unit who were somewhat peculiar that we had not 'tied' on the free end—it had disappeared back up into the abdominal cavity. He grew Escherichia coli from the cerebrospinal fluid and

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