dramatic improvement in the clinical state and the motor nerve conduction velocity would not be surprising. It is unlikely that any damage already caused by the excessive sulphatide would be corrected and the most one could hope for with the low vitamin A therapy would be to reduce any further accumulation of sulphatide and thereby limit the progression of the disease.

Early diagnosis is therefore all important. Peripheral motor nerve conduction velocity measurement (Fullerton, 1964; Aziz and Pearce, 1968) could be useful in this regard as a screening test, especially in the early stage when the only abnormal signs may be a slight hypotonia, or a genu valgum deformity with depressed or absent tendon reflexes. The diagnosis should thus be suspected in any previously normal child developing hypotonia or delay in motor milestones in the 2nd year of life. If a slow motor nerve conduction velocity is found the diagnosis can then be confirmed by demonstrating low levels of arylsulphatase activity in the blood or urine (Percy and Brady, 1968; Austin, McAfee, and Shearer, 1965). In our family we have also been able to show some reduction in levels of arylsulphatase A in the blood and the urine in the parents as well as the younger sib. It is thus possible also to identify heterozygotes for the recessive gene.

While it is difficult to assess the value of any form of therapy in a slowly progressive condition on the basis of a single case, we have been sufficiently impressed by the change in this patient to draw attention to its possible value, and at the same time to stress the importance of early recognition of this condition for future therapeutic trials.

Summary

A 3-year-old girl with metachromatic leucodystrophy showed an apparent improvement when put on a diet low in vitamin A. During a 2-year follow-up she subsequently showed no progression of the disease. The importance of early diagnosis by nerve conduction velocity studies and measurement of arylsulphatase levels in the leucocytes or urine is stressed in order to try and arrest the disease at an early stage. Heterozygotes can also be identified by lowered arylsulphatase levels in leucocytes and urine.

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Simple Method for Cutting Transverse Sections of Hair

Comments on Shape of Hair in Hurler and Sanfilippo Syndromes

In past reports on the mucopolysaccharidoses the hair has been described as coarse, but no detailed studies have been made. This paper presents a simple method of preparing transverse sections of hair for microscopical examination, and describes some features of the hair of patients with the Hurler and Sanfilippo mucopolysaccharidosis syndromes.

Materials and Methods

Hair samples were collected from the crown of the head and cut at scalp level. Samples were obtained from 2 patients with Hurler's syndrome, 5 patients with the Sanfilippo syndrome, and 10 randomly chosen normal people. Subsequently, 150 patients and staff of the Royal Children's Hospital were examined and

samples were obtained from 11 with hair which felt coarse to the fingers. The samples were sectioned transversely using a technique modified from that of Ford and Simmens (1959).

A stainless steel plate (7·4 cm × 2·2 cm × 26 S.W.G.) was used, with four holes 1 mm in diameter drilled along the centre line. The holes were contoured at each end. The plate was finished to a high polish and made perfectly flat. A loop of pull-through yarn (40 gauge (cotton or nylon) was passed through the hole. Six packing threads (150 denier cellulose acetate yarn*) and a small bunch of hair fibres were inserted into the loop. The loop was pulled through the hole, with the packing threads and hair fibres. Additional packing threads were added to the bundle and pulled through the hole until the fibres were held tightly in the hole. The bundle was then cut flush with each side of the plate using a sharp flexible razor blade, tilted at an angle of 35° to the surface of the plate and flexed gently against it to reduce slightly the angle of the cutting surface. A slicing action was used. These procedures are shown diagrammatically in Fig. 1.

The sections were observed and photographed in the plate. Thin sections were made of some samples by placing a drop of 2% cellulose acetate in acetone over the packed hole. When dry the thin film was sliced off with a razor blade. The film containing the thin sections (approximately 50 μ) was trimmed and mounted in DPX. The thin sectioning procedure could be repeated 5 to 6 times, to give serial sections. Histological staining can be performed on sections in the plate.

The shape of transverse sections of hair fibres from each sample was observed and the percentage of triangular shaped hairs (three more or less distinct sides) was counted.

*Courtaulds Aust. Ltd., Melbourne.

Results

The appearance of normal hair in transverse section is shown in Fig. 2. The round oval shape of individual shafts contrasts with the variable shape seen in hairs from a patient with Sanfilippo syndrome shown in Fig. 3. The percentage of triangular hairs in each of the samples observed is shown in the Table.

Triangular hairs were found in only 3 of the 10 normal samples (7%, 9%, 25%). These samples felt coarse. 6 of the 11 patients, chosen because their hair felt coarse, showed triangular hairs (3–36%).

Of the 12 samples from mucopolysaccharidosis patients, 10 contained triangular hairs. The 2 exceptions were samples from children aged 1 year and 2 years respectively, and later samples from both children showed triangular hairs.

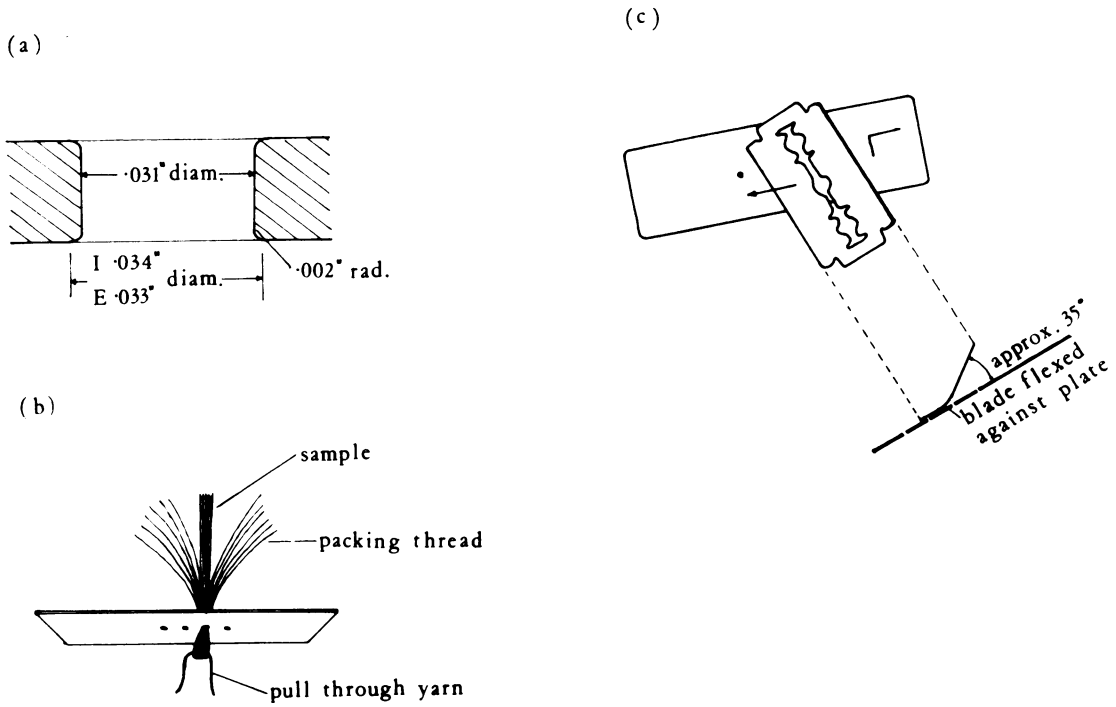

FIG. 1.—*Technique described in text. (a) The hole in the plate. (b) Packing. (c) Section cutting.*

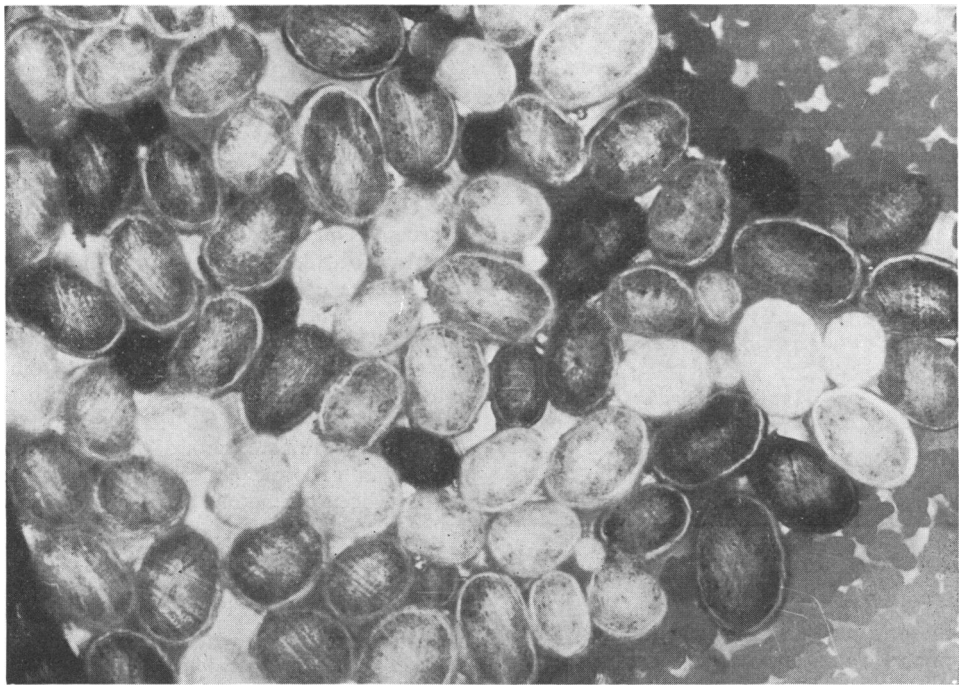

FIG. 2.—*Unstained transverse section of normal hair photographed in the plate.* (× 160.) *The hairs are round or oval.*

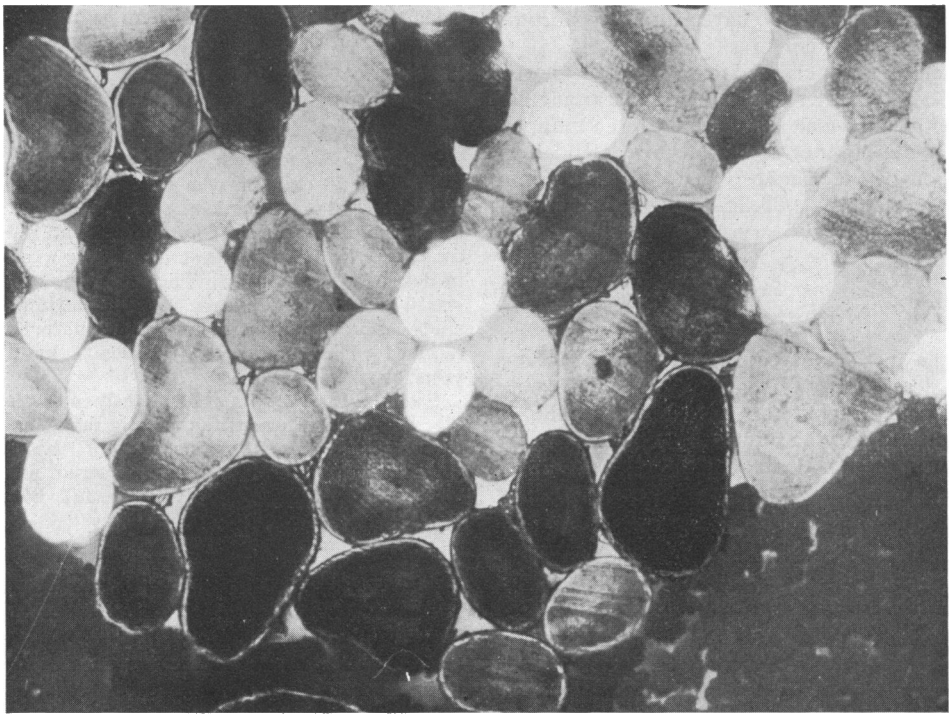

FIG. 3.—*Unstained transverse section of hair from patient with Sanfilippo syndrome.* (× *170.*) *The hairs are of variable shape, some being triangular.*

TABLE

Frequency of Triangular Hairs in Patients with Hurler and Sanfilippo Syndromes

Patient (Sex)	Age (yr)	Total No. of Hairs	% Triangular
Hurler syndrome			
S.A. (F)	9½	49	26·5
	10	56	16·1
S.C. (M)	1	80	0·0
	2	100	6·0
Sanfilippo syndrome			
D.W. (M)	12	48	29·2
	14	60	13·3
M.L. (F)	6	60	26·7
	7	83	12·0
G.S. (F)	9½	73	19·2
	10	38	26·3
W.T. (M)	4	26	15·4
	5	30	26·7
P.T.	2	32	0·0
	3	44	4·5

Discussion

From these results it seems that the presence of triangular hairs is one characteristic that contributes to a palpable coarseness of normal human hair. Triangular hairs are frequently seen in patients over the age of 2 years who have Hurler's syndrome or Sanfilippo syndrome.

This finding is not claimed to have any diagnostic value. Should further studies reveal that triangular hairs are a universal feature of the Sanfilippo syndrome, the absence of this feature could help in exclusion of the diagnosis in retarded children with coarse features. Clinical recognition of this syndrome is not always easy, even in children 5 to 10 years of age.

Summary

A simple method of cutting transverse sections of hair is described. Triangular-shaped hairs were found in patients with the Hurler and Sanfilippo syndromes and may account for the coarse feeling of the hair in these patients. Triangular hairs were also found in normal persons with coarse feeling hair.

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Value of Blood Phenytoin Estimation in Management of Childhood Epilepsy

Phenytoin has been invaluable in the management of epilepsy for over 30 years. It has many side effects, but serious adverse reactions are very unusual. Optimum control of therapy is difficult, however, because of a low toxic/therapeutic ratio and because of variations between patients in the rate of metabolic destruction of phenytoin (Kutt and McDowell, 1968). Many drugs, including several anticonvulsants, alter phenytoin metabolism and may lead to intoxication with the drug (Taylor, 1970).

In an attempt to obtain optimum control of therapy in epileptic children receiving phenytoin we have been estimating blood phenytoin levels.

Method

A modification of the method described by Dill *et al.* (1956) was employed. Phenytoin is extracted from blood into an organic solvent, evaporated, and the residue nitrated. The nitro compound is reduced to form an aromatic amine, is diazotized, and finally coupled with N-I-napthylenediamine dihydrochloride to give a coloured solution, the optical density of which is then read at 550 nm. Standard preparations were prepared from sequestrated blood. Citrated whole blood obtained from blood transfusion gave low optical densities, and was not used for this purpose.

A 2·5 ml specimen of sequestrated blood was required from the patient for each estimation.

The first 30 children who attended as outpatients or were admitted to the ward and were at that time receiving phenytoin alone, or in a combination with phenobarbitone, were included in the study. They were observed in detail for six months. The age range of the group was 6 months to 12 years. At the beginning of the study period the length of time the patients had previously received phenytoin ranged between 1 month and 5 years.

Each patient was seen at fortnightly intervals until the blood levels had neared therapeutic range, and then at monthly intervals. At the end of the six-month observation period each patient has serum folate, blood sugar, liver function tests, and a full blood count performed.

Results

The striking finding on initial investigation was the fact that only 25% of the children had therapeutic blood levels. Fig. 1 and 2 show the blood levels of the first 20 patients at the beginning and the end of the survey. At the end of the survey, 80% of the children had therapeutic levels.

Within the group, no blood abnormalities or folic