Correspondence

Arteriohepatic dysplasia

SIR,

We were interested by the article recently published by G. H. Watson and V. Miller (Archives, 1973), as since 1956 we have observed almost similar facts. We studied our patients essentially for their chronic cholestasis. Among 30 children with hepatic ductular hypoplasia, 15 comprised a distinct, homogeneous, readily recognizable group (Alagille, Habib, and Thomassin, 1969; Alagille and Thomassin, 1970). This new syndrome is more complex than proposed under the 'arteriohepatic dysplasia' label.

The association of chronic cholestasis related to hepatic ductular hypoplasia (15/15), characteristic facies (15/15), and pulmonary arterial stenosis (11/15) is almost always present in this group. But other abnormalities are almost as frequent: vertebral arch defects (8/15), growth retardation (8/15), mental retardation (9/12), hypogonadism in males (6/7) with spermatogenic hypoplasia in the 5 testicular biopsies performed.

Watson and Miller probably emphasized the pulmonary arterial stenosis because they are interested in the field of heart diseases. However, we disagree with this point, as 2 of our 15 patients had no heart abnormality, and 2 others had a different abnormality. We also disagree with their suggestion about genetic transmission: in our group there was a family history in sibs for 5 of the 15 patients, but never in parents (Alagille et al., 1968). This is why we suggest, as they do, the possibility of a genetic disorder which could either have autosomal recessive transmission or be due to chromosome abnormality. This last possibility is suggested by associated multiple congenital malformations. However, chromosomal and dermatoglyphic studies in our patients have yielded normal results. Nevertheless, infections or toxic agents transmitted in utero from the mother to one or more offspring may also produce the teratogenic effects observed.

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References


Dr. Watson replies as follows:

We are glad to comment on the letter from Professor Alagille, which clearly relates to the same syndrome as

Urinary diversion in children

Sir,

It is now 20 years since I performed my first operation for urinary deviation in a child. As the years passed I became less enthusiastic about the operation and, generally speaking, I prefer a colonic conduit to an ileal conduit. I can now present to Dr. S. R. Meadow (Archives, 1973, 48, 657) a series of girls aged 8 to 12 years with normal upper urinary tracts whose neuro-pathic bladders have been emptied by manual expression. Cine-radiography has shown that reflux has not resulted from this manoeuvre.

With the enthusiastic co-operation of the parents, I propose to continue treatment of many of my patients in this way until Professor John Lenihan (Medical Physics and Bio-Engineering, University of Glasgow) can provide my patients with a suitable bladder pace-maker.

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References


Oleic acid increased the unconjugated bilirubin in the liver slices.

I cannot agree that it is only breast milks from mothers of children with breast milk jaundice which develop increased inhibition to bilirubin conjugation on storage. The original observation (Hargreaves, 1970) that this could occur in milk from mothers of nonjaundiced children has been confirmed (Bevan and Holton, 1972). These authors also showed that the increased activity was associated with an increase in the free fatty acid content of the milk. The observation was thought to have practical implications in that deep-frozen milk is used for feeding premature infants, but it was shown that the inhibitory activity does not develop in sterilized milk (Hargreaves, 1970).

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Urinary diversion in children

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