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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REFERENCES

Urinary diversion in children

Sir,

It is now 20 years since I performed my first operation for urinary diversion 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 nephro-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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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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Dr. Watson replies as follows:

We are glad to comment on the letter from Professor Alagille, which clearly relates to the same syndrome as
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that described by us, and to see that the delineation of this syndrome has received such eminent support. Professor Alagille has very kindly sent us reprints of his publications, in which he notes the frequency of systolic murmurs in his cases of intrahepatic biliary hypoplasia (7 in 25) and which contain facial photographs of 3 of his cases who clearly resemble some of ours. His letter completes the picture by revealing the concurrence of the facial and cardiovascular features in some of his patients with liver disease, and by indicating that their murmurs are commonly due to pulmonary artery stenoses. We feel that the latter point helps to justify the name we used to epitomize succinctly the chief, though not of course the only, features, even though not all cases may have both main manifestations.

As far as the facies is concerned we feel that, though it may be characteristic in some cases, in others the features are more elusive. The other defects in Professor Alagille’s cases are not dissimilar to ours except for the mental retardation: all our schoolchildren attended normal schools, though one younger child may now turn out to be retarded.

In 3 of our families there seemed little doubt that one parent had signs compatible with pulmonary artery stenosis, and these 3 parents had an odd facies with some features of the syndrome. As two of these parents were fathers, we felt that maternal transmission of some toxic or infective agent was improbable.

We must apologize to Professor Alagille and his team for not having been able to refer to their work, but unfortunately his relevant article was published first in a periodical not quoted by the Index Medicus or Excerpta Medica; and there is no hint of any extrahepatic features in the summary at the end or in the available precis in English of the second publication of his article.

Dr. D. M. Danks also writes from Melbourne telling us of a number of his patients who have similar features to those of our patients with intrahepatic biliary atresia, and mentioning an article by Porter, Soper, and Tidrick (Annals of Surgery, 1968, 167, 602) describing 2 cases which probably also have the same syndrome.

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