Correspondence

Archives of Disease in Childhood, 1973, 48, 984.

Wilms’s tumour—a correction and an apology

Sir,

My faith in the flawlessness of British medical writing has been shattered by the repeated misspellings of Professor Wilms’s name in the paper by Evans and Holzel (Archives 1973, 48, 645).

Not only was poor old Professor Wilma’s name misspelt throughout the body of the paper, but even more disrespectful to his memory was the diligent effort on the part of someone to change the correct spelling of Wilms’s surname to the incorrect one in all the references cited in which Wilms’s name is given.

Please restore my faith in medical editing on your side of the Atlantic.

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Dr. Cone is of course correct, and we apologize abjectly. We will try to do better in future, and to restore Dr. Cone’s faith. Editors.

Effect of fatty acids on bilirubin conjugation

Sir,

We were interested in the article by T. Hargreaves (Archives, 1973) concerning the interaction between fatty acids and bilirubin conjugation in vitro. In an earlier study (Levillain et al., 1972) we showed similar results with unsaturated fatty acids but also some inhibition of conjugation with saturated fatty acids. The discrepancy between these results could be due not only to a different concentration in incubation milieu (we used approximately a concentration of 2 mmol/l. for each saturated fatty acid), but essentially to a different relative concentration of fatty acid, bilirubin, and albumin. This possibility was tested in a recent study (Luzeau et al., 1973a).

We also found (Levillain et al., 1972; Luzeau et al., 1973a) a relation between the inhibitory effect of mother’s milk samples on the glucuronyl transferase activity in vitro and their concentration of free fatty acids. Milk samples from mothers whose infants had a prolonged neonatal jaundice inhibited the enzyme activity and contained more free fatty acids than control samples. This difference cannot be detected in fresh milk samples but becomes obvious after storage, depending on duration and temperature conditions. An important liberation of fatty acids only takes place in pathological milks when they are kept at +4 °C or +20 °C while their in vitro inhibitory effect increases. Such liberation does not take place when the milk samples are preheated at +56 °C for 15 min immediately after the collection. The results obtained suggest the existence of an abnormal lipolytic activity in the milk of mothers whose infants have a prolonged jaundice. Lipoprotein lipase seems to be the enzyme responsible for the activity. These experimental data allowed us to feed 3 icteric infants with maternal milk after heating at 56 °C for 15 min: 3 or 4 days later the jaundice had disappeared. A simplified method for detection of these inhibitory milk samples has been recently described using nile blue reaction (Luzeau et al., 1973b).

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REFERENCES


Dr. Hargreaves comments as follows:

The letter from Professor Odèvre and his colleagues emphasizes yet again the difficulties in elucidating the cause of breast milk jaundice (Arias et al., 1964; Hargreaves and Piper, 1971). The observed differences between the effects of fatty acids are probably due to the concentrations used. I used a maximum concentration of 1 mmol/l. for the investigations of saturated fatty acids on bilirubin conjugation in rat liver slices (Archives, 1973, 48, 446 whereas Levillain et al. (1972) and Bevan and Holton (1972) used higher concentrations. In rat liver microsomes I found no inhibition of bilirubin conjugation by saturated fatty acids even at high concentrations. Bevan and Holton (1972) compared the effect of inhibitory breast milk and oleic acid on the bilirubin content of rat liver slices. They showed that the effect was similar except that high concentrations of
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 teratogenetic 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...