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Wolman Disease: Suggestions for Effective Treatment

For over a year a female infant affected by Wolman disease and treated by the dietary treatment described below survived far beyond the 3-4 months predictable life span. Although otherwise normal, with time she developed EFA (essential fatty acid) deficiency which can be, and is now, treated by an established procedure which is not antagonistic to the dietary treatment.

The treatment consists of strict cessation of breast feeding and strict avoidance of foods containing any fats and oils (triglycerides and cholesterolesters). Formula feeding should be as free as possible of neutral lipids, but should contain all vitamins.

In order to avoid dermatological complications and stunted growth due to EFA deficiency, about 10 microliter (1/100 of ml) of sunflower oil should be lightly rubbed, once daily on the skin of one arm in infants below 5 kg in body weight. Infants 5-10 kg in weight should be treated by

daily doses of 20-25 ml. In bigger children the dose should be about 2 mg/day/kg body weight. The site of oil smearing should be alternated on consecutive days between arms, forearms, thighs and legs.

At present it is not known whether this percutaneous administration of EFA should continue uninterrupted, or if 3-4 weeks of treatment should be followed by intermission of 1 or more weeks. It is proposed that this point should be tested by experience in different cases.

In order to be able to learn about best treatment modalities from a sizable group of patients, it is suggested that treatment of patients and its effects be reported to me every 3-6 months. A report in the name of all participants (after consultation, of course) will be published in due time. Alternately, pediatricians of a country or an area can combine their efforts and results and publish them independently. Also in this case, I would be grateful for information about the cases, which information will be considered confidential.

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Chloramphenicol Resistant S. typhi

In one(1) of the recent articles(1-3) on resistance of *S. typhi* to chloramphenicol and other drugs, antibiotic sensitivity was done by Stokes method(4). The authors of other two articles have stated that disc diffusion method was used.

It is well known that disc diffusion method is influenced by many factors like depth of medium, pH, certain cations, supplements and amount of inoculum, etc.(5). Without use of control strains on the same plate as in Stokes method(4), the results and interpretation of the results are unreliable and tend to be subjective.

In most of the laboratories, including many teaching hospitals the antibiotic sensitivity is done without use of proper control strains. The reporting is entirely on the subjective impression rather than the comparison with known standard strains. Hence, it would be advisable for authors to ensure that standard methods are used and mention in the article about it. The ideal method would be the MIC(6).

Unless standard methods are insisted upon, the results are likely to give a false and alarming picture of resistance of micro-organisms to antibiotics.

B. Subramanyam, C.S. Lakshminarayana.

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Reply

We are thankful for the interest shown in our article(1). We share the apprehension expressed by Drs. Subramanyam and Lakshminarayana that antibiotic sensitivity done by disc diffusion method (Kirby-Bauer technique) is subject to performance errors and unreliable interpretations.

In our study, the antibiotic sensitivity of Salmonella strains were done by Stokes technique on Mueller-Hinton agar with standardized inoculum(2). Simultaneously, known sensitive strains of NCTC Eschrichia coli were used as control. This facilitated standard comparisons and we never encountered any difficulty either in performance of this test or in its interpretation.

The therapeutic response to chloramphenicol correlated well with antibiotic sensitivity pattern. All patients infected with chloramphenicol sensitive S. typhi responded well to this drug and none infected with resistant strains responded to chloramphenicol.

We endorse the view that MIC is superior to all other techniques for antibiotic sensitivity. However, technically being more elaborate, it may not be feasible to use this technique as a routine procedure in busy laboratories.