

was continued in two patients (cases 1 and 2) for this reason.

Poisoning may result if the method of administering Eumydrin (as a specific number of drops) is not understood⁶ and this should be stressed whenever it is prescribed.

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Fat loss during feeding of human milk

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SUMMARY A rise in the fat concentration of human milk within the syringe was noted towards the end of continuous infusion but not with intermittent bolus gastric feeding. The rise in the former was reduced most simply and effectively by using an eccentric nozzle syringe and tilting the pump up at an angle of between 25° and 40°.

Certain methods of feeding of human milk such as continuous infusion with a syringe are associated with a rise in fat concentration towards the end of the feed.¹ We have also observed that some pumps retain this energy rich terminal milk in the syringe. We compared the fat concentration at the end of two types of enteral feeding and evaluated some methods of avoiding an excessive rise in this during infusion of human milk.

Material and methods

The fat in milk samples was estimated by the creatatocrit method² at the beginning and at the end of two types of feeding—continuous infusion using a syringe pump and intermittent bolus gastric feeding. It was observed that while some pumps emptied completely at the end of a feed there was retention of about 2 ml milk in the barrel of others. To estimate the fat concentration towards the end of

the feed the pumps were stopped when 2 ml milk remained in the syringe. Infusion feeding was studied using central and eccentric nozzle syringes and by alternative techniques detailed in the first column in the Table. Preliminary investigations showed that with the eccentric nozzle syringe fat retention was greater with the pump sloped up at angles less than 17° and more than 45°. Further investigations were therefore limited to positions within that range. In intermittent bolus gastric feeding the syringe was detached from the feeding tube when 2 ml milk remained in the barrel. All continuous infusions lasted between 3 and 4 hours and intermittent bolus feeding between 5 and 15 minutes. The results were subjected to standard statistical tests.

Results

The fat concentrations at the beginning and end of the feed and the difference between these values (mean (SD)) for the various methods are shown in the Table. There was no rise with intermittent bolus feeding (method (a)) and even when a sample of milk was aspirated from the feeding tube at the end of a feed fat concentration was not increased.

With the central nozzle syringe the rise in fat concentration was highest in the conventional method with the pump kept horizontal (method (b)1), increasing to a maximum of 40%. When some milk was aspirated from the tube the concentration

Table Variations in fat concentration during feeding of human milk (estimated by creatocrit method²; values percentage mean (SD))

Method	No of studies	Initial concentration	Final concentration	Difference
(a) Intermittent bolus feeding	50	6.3 (1.7)	5.9 (1.7)	-0.3 (0.5)
(b) Continuous infusion				
Central nozzle syringe				
1 Horizontal	50	5.4 (2.4)	26.8 (5.3)	21.4 (6.0)
2 Vertical	10	3.7 (0.8)	16.2 (3.2)	12.5 (3.5)
3 Horizontal with hourly mixing by syringe agitation	10	4.8 (1.4)	26.7 (2.7)	21.9 (2.0)
4 Horizontal with half hourly mixing	10	5.5 (1.5)	10.2 (1.2)	4.6 (0.7)
5 Vertical with hourly mixing	10	4.9 (0.9)	9.7 (0.6)	4.8 (0.6)
Eccentric nozzle syringe				
6 Horizontal	10	4.2 (1.1)	17.0 (2.6)	12.8 (2.1)
7 Horizontal with hourly mixing	10	4.5 (1.0)	11.1 (2.2)	6.6 (1.5)
8 Oblique—17°	10	4.6 (0.8)	12.2 (2.0)	8.0 (1.7)
9 Oblique—25°	10	4.8 (1.0)	7.3 (1.6)	2.5 (1.3)
10 Oblique—37°	10	3.8 (0.8)	7.1 (1.5)	3.3 (1.0)
11 Oblique—40°	10	4.1 (1.0)	7.2 (1.5)	3.1 (1.1)
12 Oblique—45°	10	3.2 (1.1)	11.9 (2.6)	8.7 (1.8)

was at times as high as 50%. With the exception of method (b)3 (that is hourly agitation of the syringe) terminal fat concentration was significantly lower with other methods such as (b)2 and 6 (Table) ($P < 0.01$), even lower with the alternative techniques listed, and was least with an eccentric nozzle syringe tilted up at an angle of 25° to 40° (methods (b)9, 10, and 11). Accidentally, the eccentric nozzle syringe was placed with the nozzle tilted up but on the lower side of the barrel on two occasions, and for obvious reasons the rise in the fat at the end of these feeds was extremely high—40% and 45% respectively.

A prototype wooden wedge (Figure) was constructed by the local engineering department with a

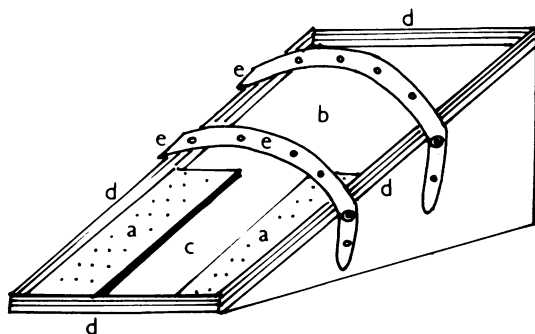


Figure The ramp used in the study for supporting the infusion pump in an oblique position.

This was broad enough to accommodate different types of pumps. Two strips of wood fixed on either side (dotted areas (a)) allowed placement of wider pumps with back projections on part (b). Long narrow pumps could rest in the shallow trough (c). A raised ridge (d) at the periphery prevented the pumps from sliding off. Further stability was achieved with rubber straps (e) fixed to hooks on the side of the ramp.

ramp at an angle of 25° to the horizontal; this narrower angle was selected to make the wedge more stable. The ramp was tested in the unit and no problems such as leaking of milk from the back of the syringe were noted by the nurses. Central nozzle syringes were also tried on this ramp as currently available eccentric nozzle syringes fitted into only some of the infusion pumps. The percentage rise in fat with the latter was mean (SD) 12.1 (3.6), which was greater than with the eccentric nozzle syringe in the same position ($P < 0.01$), but less than with the central nozzle syringe in the conventional horizontal position ($P < 0.01$).

Discussion

Separation of fat in human milk during continuous infusion may contribute to an appreciable fat loss during prolonged periods of feeding, especially if some of the fat rich milk is left in the syringe at the end of each feed. One method of avoiding this would be to homogenise the milk but it is preferable to minimise processing human milk so that its unique properties are retained better.

A simple way to reduce fat loss is to ensure that the syringe is completely emptied at the end of each feed. With the conventional infusion method, however, terminal fat concentration can be excessive—up to 40% or more. While there is some rise in the milk fat concentration towards the end of normal breast feeding which is physiological,^{3,4} the rise during conventional infusion seems to be abnormal. In fact, it would be almost equivalent to introducing a small pat of butter every 3 to 4 hours and could not be physiological. Periodic agitation of the syringe to keep the fat concentration more uniform can be carried out but it is time consuming

and has the potential risk of causing separation of solid fat (that is, butter). Finally, since fat is lighter its expulsion is aided by the use of an eccentric nozzle syringe, especially if the nozzle is tilted up as is done to expel air bubbles. Even in the horizontal position the eccentric nozzle syringe seems to be better. If the syringe and pump are tilted up care must always be taken to fix them well to ensure stability and to avoid accidental fall. Additional suggestions include the use of shorter feeding and connecting tubes, complete emptying of the syringe at the end of each feed, and the early initiation of intermittent bolus feeding. It would also be beneficial if manufacturers of infusion pumps and syringes ensured a basic uniformity of design to allow easy fitting.

It is important to make sure that all the nutrients in human milk are actually delivered to the infant.

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Severe ornithine transcarbamylase deficiency

Two and a half years' survival with normal development

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SUMMARY The clinical course and management of a boy with severe ornithine transcarbamylase deficiency are described. In addition to treatment with sodium benzoate and amino acid keto analogues, mannitol may be useful in hyperammonaemia and nocturnal gavage feeding aids maintenance treatment.

Untreated, severe ornithine transcarbamylase deficiency causes fatal neonatal hyperammonaemia. Treatment prolongs survival but only two boys living more than 18 months have been reported—one aged 22 months with normal development and one 5 year old with severe psychomotor retardation.^{1 2} We report a third.

Case report

A boy born in June 1981, the fourth child of unrelated French parents, was transferred to this hospital aged 3 days. His brothers had died as

neonates—two from congenital malformations with no hyperammonaemia and one from ornithine transcarbamylase deficiency. His mother is asymptomatic but her serum ammonia and urine orotic acid concentrations rose abnormally on protein challenge. Jejunal biopsies in both parents showed normal ornithine transcarbamylase activity.

On hospital admission the boy was unresponsive with a serum ammonia value of 714 $\mu\text{mol/l}$ (normal value less than 60 $\mu\text{mol/l}$) and he had *Escherichia coli* septicaemia. Intravenous antibiotics and a dietary mixture given by gavage stopped deterioration, but on day 7 osmotic diarrhoea led to dehydration and coma and his serum ammonia value was 819 $\mu\text{mol/l}$. He improved with exchange transfusion, diluted feeds, and sodium benzoate and was clinically normal on day 10. After milk feeding was started on day 18 he thrived and his height and growth centiles were -2 SD and -1 SD respectively.

He has had three febrile illnesses and three immunisations without problem. At age 3 months he had a needle liver biopsy. At age 19 months he had a