

CYSTINOSIS MEDICAL ALERT PAGE FOR PATIENTS AND FAMILIES

Children with cystinosis are particularly prone to the effects of dehydration and their carers and physicians need to be fully aware of the risks for them. This alert page is to guide families and their doctors in order that these children receive proper assessment and appropriate treatment.

All children with the following symptoms need an assessment by a medical practitioner:

- Persistent vomiting
- Diarrhoea
- Diarrhoea with vomiting
- Vomiting with fever

All children with Cystinosis who lose fluid from their body, either through diarrhoea or vomiting, are at risk of dehydration. This occurs more quickly than in other children. Thus these children need early review. In addition, these children are dependent on the electrolyte supplements that they have been prescribed and a short period of vomiting can have a rapid effect on the blood electrolytes if the medication cannot be given regularly as prescribed.

The doctor must:

- Assess the child's hydration status
- Check the weight
- Compare the weight with the previous stable weight (usually the family knows the child's most recent weight)
- Arrange a blood test to check electrolytes, in particular to look for changes in blood sodium, low potassium and acidosis

Changes in the electrolytes as well as the development of acidosis can be very rapid and will require establishment of fluid replacement either through nasogastric tube or gastrostomy if the child can tolerate this orally or via intravenous fluid. The choice of intravenous fluid will depend on the blood sodium concentration but a standard replacement prior to obtaining the results would be 0.45 saline, 5% dextrose with 20 mmols potassium chloride per 500 ml bag of solution. Acidosis will need to be treated appropriately with sodium bicarbonate or potassium citrate.

Please always check with the child's specialist paediatrician if in doubt.