Child Growth Foundation

Treatment of the SRS/SGA Child: Guidelines for Emergency Services and Inpatient Care Teams

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INTRODUCTION

This document has been prepared to assist you in the emergency care and the treatment of high risk infants and children with Silver Russell Syndrome (SRS) and/or children with non-syndromic gestational growth restriction who were born small for gestational age (SGA). It is suggested that the parents of SRS/SGA infants and children carry this document with them at all times and present it to paramedics and the inpatient team who will be caring for their infant or child. This information is based upon the authors' considerable experience of caring for hundreds of SRS/SGA children.

OVERVIEW

To efficiently care for this child in an emergency or special medical situation, you will need to consider some issues not commonly encountered and to arrange treatment that may differ from what would be routine inpatient management for a normal child with a similar illness.

This group of infants and children are at an especially high risk of developing fasting hypoglycaemia and starvation ketosis for the following reasons:

- They frequently have feeding difficulties, gut dysmotility and an abnormal appetite which causes intolerance or sufficient enteral calories when ill or stressed.
- They have a small liver and a low muscle mass that limit glycogen storage.
- Their small liver and low muscle mass restricts both their capacity for and their amino acid substrate for gluconeogenesis.
- They have a normal-for-brain age but an abnormally small-for-age body, which increases the need for glucose substrate but limits storage and maintenance of glucose substrate.
- They frequently have growth hormone insufficiency, which further impairs their ability to maintain a normal circulating glucose through gluconeogenesis.

Since these childrens' hypoglycaemia is associated with a lot glycogen state, glucagon is contraindicated. Their impaired fasting glucose homeostasis combined with their endogenous insulin resistance causes a risk of both fasting hypoglycaemia and fed hyperglycaemia.

FOR THE EMERGENCY SERVICES

To quickly resuscitate these children, we suggest the following:

- Test the blood sugar as you start an IV.
- Include the equivalent of 10% dextrose in the IV infused at a maintenance fluid rate, if the blood sugar is less than 150 mg/dl.
- Avoid a glucose bolus if possible.
- Check every urine for ketones.
- Follow finger stick blood glucose hourly until stable.
- Weigh without a nappy or in underwear on admission.
- Consider the child's size not their age for all calculations.
- Follow intake and output carefully.
- Keep NPO if vomiting.
- Treat fever aggressively.
- Arrange for an admission to the hospital if vomiting, febrile or spilling ketones and not tolerating full feeds or is hypoglycaemic.
- Weigh on discharge.

- Pass this document to the inpatient team.

Because of their generalised gut dysmotility and lack of a normal appetite, SRS/SGA children can be very difficult to re-feed after an acute illness, especially if associated with vomiting. Because their normal PO/GT intake is frequently marginal, they must be feeding normally before IV carbohydrate intake can be completely discontinued.

FOR INPATIENT CARE TEAM

To effectively return these children to their normal status and avoid a rebound back to the hospital, we suggest the following:

- Weigh without a nappy or in underwear on admission and continue to weigh daily on the same scale.
- Follow intake and output carefully.
- Follow finger stick blood glucose 2 hourly until stable and after changes are made to carbohydrate infusion and PO/GT intake.
- Check every urine for ketones.
- Continue the equivalent of 2 times maintenance dextrose in an IV delivering routine maintenance fluid and electrolyte as long as patient in NPO and blood sugar is less than 120 mg/dl.
- Keep NPO for a full 24 hours after vomiting stops.
- Start enteral challenge with small volumes and advance PO/GT feeding very slowly for both volume and concentration.
- Wait 12 hours to re-challenge if vomiting recurs.
- Discharge after stable on full feeds for 12 hours without positive urine ketones.

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This document is an excerpt from the book RSS/SGA – A Comprehensive Guide: Understanding Aspects of Children Diagnosed with Russell-Silver Syndrome or Born Small for Gestational Age. Co-authored by an international group of medical professionals in fields ranging from endocrinology and paediatrics to feeding and adrenal disorders. This guidebook is a comprehensive source of information for medical practitioners, parents and caregivers of SRS/SGA children. This 330-page guidebook is available through The Magic Foundation in the USA and the Child Growth Foundation in the UK.

We thank the Magic Foundation for allowing us to reproduce this document.

FURTHER INFORMATION

If you have any questions regarding the information contained in this sheet, then please contact our Patient Support Line:

Tel: 0208 995 0257 | Email: info@childgrowthfoundation.org

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FEEDBACK

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