Treatment of the Ill RSS/SGA Child: Guidelines for First Responders, Emergency Services and Inpatient Care Teams
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Introduction
We have prepared this document to assist you in the emergency care and the treatment of a special group of rare, high risk infants and children with Russell-Silver Syndrome (RSS) and/or children with non-syndromic gestational growth restriction who were born small for gestational age (SGA). We have suggested that the parents of RSS/SGA infants and children carry this document with them at all times and present it to First Responders and the inpatient team who will be caring for their infant or child. This information is based upon our twenty plus years of experience caring for hundreds of RSS/SGA children. To efficiently care for this child in an emergency or special medical situation, you will need to consider some issues not commonly encountered by first responders and to arrange therapy 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 for developing fasting hypoglycemia and starvation ketosis for the following reasons:
- They frequently have feeding difficulties, gut dysmotility and an abnormal appetite which cause intolerance of 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 a low muscle mass restricts both their capacity for and their amino acid substrate for gluconeogenesis.
- They have a normal-for-age brain 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. [Not indicated]

Since these children’s hypoglycemia is associated with a low glycogen state, glucagon is contraindicated. Their impaired fasting glucose homeostasis combined with their endogenous insulin resistance causes a risk of both fasting hypoglycemia and fed hyperglycemia.

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 75 mg/dl.
- Avoid a glucose bolus if possible.
- Check every urine for ketones.
- Follow finger stick blood glucose hourly until stable.
- Weigh without a diaper or in underwear on admission to the emergency room.
- Consider the child his size not his age for all calculations.
- Follow intake and output carefully.
- Keep NPO if vomiting. ["NPO"=nothing by mouth]
- Treat fever aggressively.
- Arrange for admission to the hospital if vomiting, febrile or spilling ketones and not tolerating full feeds or is hypoglycemic.
- Weigh on discharge from the emergency room.
- Pass this document to the inpatient team.

Because of their generalized gut dysmotility and lack of a normal appetite, RSS/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.

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For the 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 diaper or in underwear on admission to the pediatric floor and continue to weigh daily on the same scale.
- Follow intake and output carefully.
- Follow finger stick blood glucose q 2 hour 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 patient is NPO and blood sugar is less than 100 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 urinary 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 pediatrics to feeding and adrenal disorders, this guidebook is a comprehensive source of information for medical practitioners, parents and caregivers of RSS/SGA children. This 330-page guidebook is available only through The MAGIC Foundation.

Fifteen chapters include topics such as:
- Physical Characteristics and Functional Abnormalities
- Fasting Hypoglycemia and Ketonemia
- Improving Your Child’s Weight
- Moving into Adulthood: A Discussion for those Born RSS/SGA

To receive a copy of the Guidebook, simply contact MAGIC at 800-3MAGIC3: any family who becomes a new member of MAGIC will receive one Guidebook free and any medical professional can request either a hardcopy or DVD free as well.

http://www.magicfoundation.org/