

Russell-Silver Syndrome at a Glance

Russell-Silver syndrome (RSS) is characterized by slow growth both in utero and after birth, short stature, and specific facial features. Children with RSS have an increased risk for developmental delay (both motor and cognitive) and learning disabilities.

RSS can be caused by changes on either chromosome 7 or 11. It occurs in about 1 in 100,000 births.

Major Characteristics

- Growth retardation (below 3rd % on growth charts)
 - The average adult height of males is ~4 feet 11 inches (151.2 cm) and that of females is 4 feet 7 inches (139.9 cm.)
 - Growth is typically proportionate
 - However, most individuals with RSS have a short arm span.
 - Slow growth in utero is an essential feature of RSS.
- Limb, body, and facial asymmetry may be present
 - Limb length asymmetry most common
 - Children may have different foot sizes
- Characteristic facial features
 - Triangular shaped face
 - Prominent forehead
 - Small, narrow chin
- Motor, speech, and/or cognitive delays

Other findings

- Hypoglycemia (low blood sugar)
 - Children with RSS have little subcutaneous fat, are quite thin, and often have poor appetites.
 - They are at risk for hypoglycemia with any prolonged fast, including surgery.
 - Excessive sweating is often associated with hypoglycemia
- Gastrointestinal findings

- Reflux
- Food aversions
- Constipation
- Skin pigment changes
 - Including café au lait spots
- Kidney problems
- Teeth crowding
 - Important to have good oral hygiene
- Fingers and toes
 - 5th finger curved
 - Short, stubby fingers and toes

1. Medical and Dietary Needs

What you need to know

The severity of any one of the possible medical conditions varies widely between individuals. Therefore it is important to ask the parents about any specific medical issues that their child may have.

School-aged children with RSS may have multiple doctors and specialist visits to monitor medical conditions.

Regular neurodevelopmental and developmental/behavioral evaluations are important.

Individuals with RSS are at increased risk of **hypoglycemia**.

- Signs of hypoglycemia
 - Excessive sweating
 - Extreme crankiness
 - Lethargy
 - Paleness and shakiness
 - Poor coordination or odd speech

A child may also have **gastrointestinal** findings:

- Reflux
- Food aversions
- Constipation

They may have food aversions.

What you can do

- Treatment for hypoglycemia
 - Dietary supplementation
 - Frequent feedings
 - Complex carbohydrates
 - Snacking during the day
 - Allow child access to snacks at all times
 - May require a gastrostomy tube if child cannot maintain blood sugars consistently
- Gastrointestinal issues
 - Keep acid blocking medications in nurse's office
 - Talk with the parents about particular foods that might be triggers for the reflux and avoid those foods if the child has more vomiting or reflux than normal.
 - If reflux is a current problem, avoid positions where the child's head is below their stomach, such as reading while lying down.
 - They may possibly have pain from constipation.
- Food Aversions
 - Individuals with RSS may require feeding therapy for food aversions
 - Speech Therapy
 - Occupational Therapy
- A yearly check-up and studies as needed should occur in the child's Medical Home.
- Be aware of any changes in behavior or mood that seem out of line with the situation and notify the parents.
- It is important to be aware of any academic changes. Contact parents when any differences are noticed.

2. Education Supports

It is important to have HIGH LEARNING EXPECTATIONS for children who have Russell-Silver syndrome. Encourage use of the core educational curriculum and modify it in order to meet the individual needs of the child.

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What you need to know

There is a wide degree of variability in cognitive and adaptive function in individuals with RSS. Learning disabilities and attention deficit disorders (ADD) appear to be increased in individuals with RSS. Autism and pervasive developmental delays (PDD) may also be increased in these individuals. It is important to have high but realistic expectations for each child.

Consider therapists and specialists to consult and support classroom teacher.

- Physical therapy
- Occupational therapy
- Speech therapy

Physical Supports

- It is important to find the balance between providing help and fostering autonomy. Individuals with RSS are able to live independent productive lives with adaptive assistance or adjustments. An IEP/504 plan may be in place for individuals' safety and comfort in the class and school.

What you can do

- Due to short stature and motor issues, adaptive aids in school may be required for:
 - Heavy doors
 - High doorknobs
 - Reaching the blackboard
 - Extenders
 - Stools
 - Desk size
 - Bathroom
 - Use nurse's office with a permanent step
 - Carrying books may be a challenge
 - Two sets of books; one for home one for school
 - Friend helper
 - Low locker
 - Allow extra time to travel between classes/use elevators

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- May need stool to rest legs on
- Occupational therapy and/or accommodations for writing
 - Individuals may have small fingers
 - May not be able to write at a quick enough speed
 - Consider tape recorder for class
 - Use computers
 - Additional time for tests or provide oral exams
- Limb length discrepancy
 - Shoe lifts may help
- It is important to treat an individual age appropriately
 - Don't baby them because they are small
 - Don't carry them
- Individuals may feel social stigma due to short stature
 - Little People Association has many resources
 - Counseling may be appropriate
 - Support groups

If Autism is a secondary diagnosis, please refer to the GEMSS autism page for further ideas and information.

3. Behavioral and Sensory Support

What you need to know

Be sensitive about body differences and short stature. This may play a role in self-image, peer relationships, and socializations.

What you can do

Consider treatments:

- Behavioral supports
- Counseling
- Medications

4. Physical Activity, Trips, Events

What you need to know

Exercise and physical education should be encouraged for strength building and obesity prevention.

What you can do

Adapting a child's physical education programs may be helpful. Here are some possible modifications, *if necessary*:

- Downsize equipment – i.e. smaller bat
- Run 2 laps instead of 4
- Have a designated runner
- If activity can't be modified to be safe, have child be a scorekeeper, umpire etc.
- Swimming and biking are encouraged
- Occupational therapy evaluation may help with accommodations and modifications
- If a field trip is planned with a lot of walking, make accommodations

If a lot of walking is planned, it will take more time and children may be tired.

- Consider cutting down on walking when possible
- Use alternative forms of transportation

5. Absences and Fatigue

What You Need to Know

Absences

- Children with RSS may be absent due to illness and/or medical appointments.

Fatigue

- Individuals with RSS should not have excessive fatigue, but consider fatigue when walking long distances is planned.

What you can do

- Help to make transitions in and out of school as seamless as possible.

6. Emergency Planning

What you need to know

- Develop an emergency plan if necessary, depending on the needs of individual children.
- See the emergency plan for hypoglycemia in RSS, from the Magic Foundation:
<http://www.magicfoundation.org/downloads/RSSEmergencyTreatment.pdf>.

7. Resources

The Magic Foundation

<http://www.magicfoundation.org>

The Magic Foundation is an organization that provides information for many conditions, including Russell-Silver syndrome.

Genetics Home Reference: Russell-Silver syndrome

<http://ghr.nlm.nih.gov/condition/russell-silver-syndrome>

Consumer-friendly information about human genetics from the U.S. National Library of Medicine

Little People's Association

<http://www.lpaonline.org/>

For resources on short stature

Note: This printable version does not include the information found under the green button marked "Transitions" on the website. Those general pages may be printed separately.