

Russell-Silver Syndrome For Healthcare Providers

This is a customized health care provider version of our website. Please visit the main website to find more comprehensive information for families and schools (www.gemssforschools.org).

Physical characteristics and/or symptoms

Russell-Silver is a condition with both intrauterine and postnatal growth deficiency. Although not all people with Russell-Silver will have all of these features, the features listed below are considered Major and Minor criteria for making the diagnosis. The diagnosis of RSS and supportive laboratory testing should be considered in individuals who have three major criteria or two major and two minor criteria.

Major Criteria:

- Intrauterine growth retardation/small for gestational age (<10th percentile)
- Postnatal growth with height/length <3rd percentile
 - The average adult height of males is ~4 feet 11 inches
 - Females is 4 feet 7 inches
- Growth is typically proportionate
- Normal head circumference (3rd-97th percentile)
- Limb, body, and/or facial asymmetry

Minor Criteria:

- Short (arm) span with normal upper- to lower-segment ratio
- Fifth finger clinodactyly (incurving of the little fingers)
- Triangular face
- Frontal bossing/prominent forehead

Supportive Criteria:

- Café au lait spots or skin pigmentary changes
- Genitourinary anomalies (cryptorchidism, hypospadias)
- Motor, speech, and/or cognitive delays
- Feeding disorder
 - Gastresophageal reflux
 - Food aversions
 - Constipation
- Hypoglycemia

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Other less common findings may include:

- Kidney problems
- Teeth crowding
- Short or curved fingers and/or toes
- Scoliosis

Recommended Routine Surveillance

- Follow growth velocity and limb length for evidence of asymmetric growth
- Monitor for hypoglycemia
- Speech and language evaluations
- Routine dental care

Emergency Protocols

There are no specific emergency protocols for this particular condition as it is not typically associated with episodes of sudden and serious medical decompensation.

- Emergencies should be handled as with any child.

Specialists Who May Be Involved

Follow up is need on a case-by-case basis. A multidisciplinary team approach to best meet the child's individual needs is recommended.

- Geneticist / Genetic Counselor:
 - Diagnosis and testing to determine the underlying genetic cause as Russell-Silver syndrome (RSS) is a genetically heterogeneous condition
 - Coordination of care
 - Genetic risk for the family
 - Clinical trials
- Gastroenterologist:
 - Feeding difficulties
 - Esophagitis
 - Gastrointestinal reflux
- Endocrinologist:
 - Hypoglycemia
 - Possible growth hormone therapy
- Orthopedist:
 - Limb length discrepancies
 - Scoliosis
- Genito-Urologist:
 - Genital abnormalities

- Developmental evaluation:
 - Speech therapy
 - Physical therapy
 - Occupational therapy

Sample Forms

Sample paragraph to be used for Letters of Medical Necessity or Letters to the school:

My patient _____ has been diagnosed with Russell-Silver syndrome. Growth deficiency and short stature characterize Russell- Sliver syndrome. Medical complications with Russell Silver syndrome include hypoglycemia, feeding difficulties, gastrointestinal reflux, and leg length discrepancies. Because of these, _____ needs the following accommodations.

Samples of paragraphs from the Little People of America to use for school letters for dwarfing conditions:

<https://lpa.memberclicks.net/assets/school%20letters%202016.pdf>

Seven Important Aspects of School Life

“[Russell Silver Syndrome at a Glance](#)” will help you talk with parents and schools about:

- Medical / Dietary Needs
- Education Supports
- Behavior & Sensory Supports
- Physical Activity, Trips, Events
- School Absences & Fatigue
- Emergency Planning
- Transitions

Resources

Genes Reviews: National Center for Biotechnology Information (NCBI) Bookshelf – Russell-Silver syndrome

<https://www.ncbi.nlm.nih.gov/books/NBK1324/>

Genetic Home Reference

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