Physician Form – Silver Russell Syndrome Screening
Using: Netchine-Harbison SRS/RSS Clinical Scoring System

Child's Name: _____   Date of Birth: _____

Gender: Male   Female   Gest Age: ____wks ____days

Father's Height: _____   Mother's Height: _____   Mid Parental Target Height (MPTH): _____

Birth Measurements – SGA Evaluation

Birth Wt:	kg	Birth Wt SDS:	SDS	SGA	Yes/No
Birth Lth:	cm	Birth Lth SDS:	SDS	SGA	Yes/No
Birth HC:	cm	Birth HC SDS:	SDS		

NH-CSS FACTORS: RESULTS

Please refer to accompanying page for definitions and notes for each scoring system factor

- *Factor 1: Being born small-for-gestational-age*   YES   NO

- *Factor 2: Relative macrocephaly at birth*   YES   NO

- *Factor 3: Postnatal growth failure ≤ -2SDS at age 24months OR
  ≤-2SDS from MPTH at 24 months*   YES   NO

- *Factor 4: Feeding difficulties &/or BMI ≤ -2SDS at 24 months* YES   NO

- *Factor 5: Protruding forehead, age 1-3yrs*   YES   NO

- *Factor 6: Body asymmetry*   YES   NO

Total Number of Factors Present in Child: _____ of 6

Score of 4-6 = Possible SRS/RSS

- Implement SRS/RSS molecular testing to investigate the known molecular SRS/RSS etiologies
- If the molecular testing cannot be performed or is negative, a child may be clinically diagnosed as SRS/RSS if differential diagnoses have been ruled out [e.g., Bloom, Fanconi, Mulibrey-Nanism, 3-M syndrome…]

Score of 3 or less = Unlikely SRS/RSS

- Using the list of other typical SRS/RSS characteristics below may be helpful in detecting the rare false negative SRS/RSS patients (a small number of mUPD7 children, or 11p15 children with tall parents, may score unlikely on the SRS/RSS scoring system).

Other SRS/RSS Physical Characteristics (each of these has been found to be statistically more significant in specific SRS/RSS groups but can also be found in non-SRS/RSS SGA/IUGR children):

____ Downturned mouth (90% of 11p15)   ____ Low muscle mass (80% of 11p15 & 70% of mUPD7)
____ Clindactyly of the 5th finger (90% of 11p15)   ____ Prominent heel (100% of mUPD7)
____ Shoulder dimples (70-75% 11p15 & mUPD7)   ____ Autism/PDD (50% of mUPD7)
____ Syndactyly of the 2/3 toes (90% of 11p15)   ____ Diagnosed cognitive disabilities (75% of mUPD7)
* incidence percentages are approximate

Netchine-Harbison SRS/RSS Clinical Scoring System - Factor Definitions and Notes

- *Factor 1: Being born small-for-gestational-age*

DEFINED: ≤-2SDS* birth length and/or weight adjusted for gestational age (GA)

* according to Usher and Mc Lean references

NOTES: The GA used should be the *first* one given to the mother and not a corrected one (often times, a due date is moved later due to the fetus measuring smaller than expected).

- *Factor 2: Relative macrocephaly at birth*

DEFINED: Birth head circumference SDS ≥1.5SDS than birth weight or length, SDS* adjusted for gestational age

* according to Usher and Mc Lean references

NOTES: Use the first available HC in first month if birth HC is unavailable or is questionable in accuracy.

- *Factor 3: Postnatal growth failure ≤ -2SDS at age 24mos or ≤-2SDS from MPTH*

DEFINED: -2SDS at/about 24 months compared to mean and/or compared to MPTH

NOTES: Make sure to note if a child is -2SDS from MPTH even if he is not -2SDS from the mean.

- *Factor 4: Feeding difficulties and/or low BMI at 24 months*

DEFINED: A BMI ≤-2SDS at 24 months OR be tube-fed OR already be on cyproheptadine for appetite stimulation

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- *Factor 5: Protruding forehead, age 1-3yrs*

DEFINED: This factor requires that the forehead protrudes from the plane of the face (defined as a forehead that projects beyond the facial plane when viewed laterally); the forehead may also be high but MUST protrude. See photo examples on back side

NOTES: For older children, scoring this factor requires looking at photos of the child between 1-3 years of age (due to fact that this factor is less evident in infancy and often disappears after age 3 years). The most typical SRS/RSS forehead is easier to diagnose due to its severity in protrusion but milder protrusion can be difficult to assess.
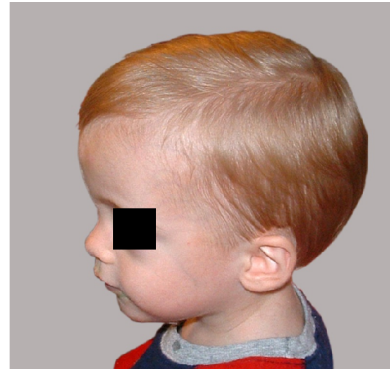

- *Factor 6: Body asymmetry*

DEFINED: a leg length discrepancy (LLD) of ≥ .5cm OR arm asymmetry OR LLD < .5cm with at least two other asymmetric body parts (with one being a non-face part)

Netchine-Harbison SRS/RSS Clinical Scoring System
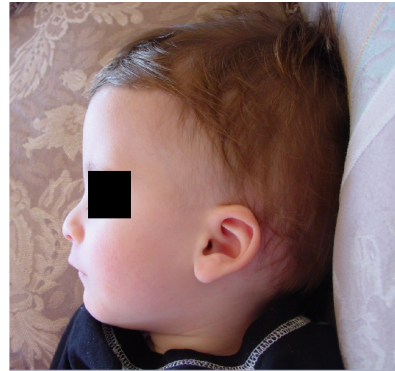## Representative Photos

Significant protruding forehead
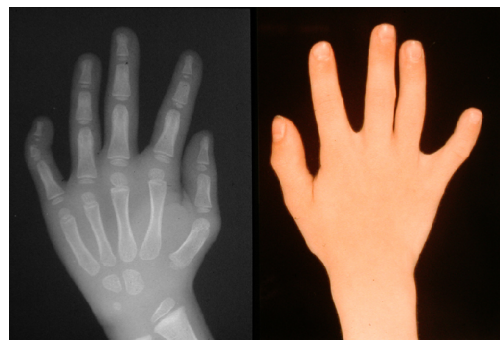
Moderate protruding forehead

Mild forehead protrusion

NO forehead protrusion but high

Prominent heel

Clinodactyly of 5th finger