OBJECTIVE

To assess the cerebral function in children with CHI at follow up.

MATERIAL AND METHODS

Study population
- International cohort
- 67 children with verified CHI
- Denmark/Sweden N=12, siblings N=5
- Eastern Europe – treated in Denmark N=22
- Russia = 33

Bayley Scale of Infant and Toddler Development, 3rd Edition (age 0-2 years = 37)
DP-3 Developmental Profile (age >2 years =20)
Wechsler’s Intelligent Scale for Children, 4rd edition (age ≥6 N=5)
Movement ABC to assess motor function (age 3-17 years, n=10).

RESULTS

Cerebral outcome for children with CHI born and treated in Russia. Tested with Bayley at age 0-2 years, with DP-3 > 2 years of age.

Eastern countries N=20

Bayley 0-2 y Percentile
Cognitive 20.3±31.6
Communication 13.1±20.3
Motor 27.7±32.0
Social-emotion 23.4±32.9
Adaptive 15.6±33.9

DP3>2 y Mean standard score
Physical 75.6±26.5
Adaptive 77.6±28.1
Social 75.3±28.6
Cognitive 70.5±26.0
Communication 73.5±25.6

Normal development ** = IQ > 90, Bayley > 40 percentile or M-ABC > 40 percentile or DP3 > 85
Moderate disabled*** =IQ: 80 – 90 Bayley or M-ABC: 15 - 40 percentile or DP3: 60 – 85.
Severe disabled ****IQ < 80 or Bayley or M-ABC < 15 percentile or DP 3 < 60.

CONCLUSION

A high degree of neurodevelopmental delay still occurs in children with CHI, in all investigated countries.

RESULTS cont.

Movement ABC-2, DK-SW

Verbal comprehension Perceptual organization Working memory

Processing speed Total IQ score

Overall outcome for children with CHI.

<table>
<thead>
<tr>
<th>Total</th>
<th>Nordic</th>
<th>Foreign</th>
<th>Russia</th>
<th>Early onset</th>
<th>Late onset</th>
<th>K-ATP</th>
<th>Other genetics</th>
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</thead>
<tbody>
<tr>
<td>N=67</td>
<td>N=12</td>
<td>N=12</td>
<td>N=32</td>
<td>N=42</td>
<td>N=25</td>
<td>N=31</td>
<td>N=36</td>
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<tr>
<td>Normal development</td>
<td>22(33)</td>
<td>7(58)</td>
<td>5(23)</td>
<td>10(31)</td>
<td>27(64)</td>
<td>13(52)</td>
<td>22(71)</td>
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<tr>
<td>Moderate disabled</td>
<td>5(7)</td>
<td>0(0)</td>
<td>0(0)</td>
<td>5(16)</td>
<td>1(3)</td>
<td>4(16)</td>
<td>1(3)</td>
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<tr>
<td>Severe disabled</td>
<td>40(60)</td>
<td>5(42)</td>
<td>17(77)</td>
<td>17(53)</td>
<td>14(33)</td>
<td>8(32)</td>
<td>8(26)</td>
</tr>
</tbody>
</table>

DP 3: delayed < 70, below average 70-84, average 85-115, above average 116-150, well above average >150.

Disclosure statement: We have nothing to declare.

Cerebral Outcome of Children with Congenital Hyperinsulinism


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