

Cerebral Outcome of Children with Congenital Hyperinsulinism

Helleskov A.(1), Melikian, M.(2), Scherderkina, I.(3), Globa, E.(4), Pørtner F.(1), Larsen AM.(1) Filipsen, K.(1), Brusgaard, K.(6), Christesen, HT.(1)

1: Hans Christian Andersen Children's Hospital, Odense University Hospital / University of Southern Denmark, Denmark. 2: Endocrinology Scientific Centre, Russian Academy of science, Moscow, Russia. 3: Morozovsky children hospital, Russian national research medical University, Moscow, Russia. 4: Department of children and adolescent Endocrinology, Ukrainian Center of Endocrine Surgery, Endocrine Organs and Tissue Transplantation, Ukraine. 5: Dept. of Clinical Genetics, Odense University Hospital, Denmark

OBJECTIVE

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

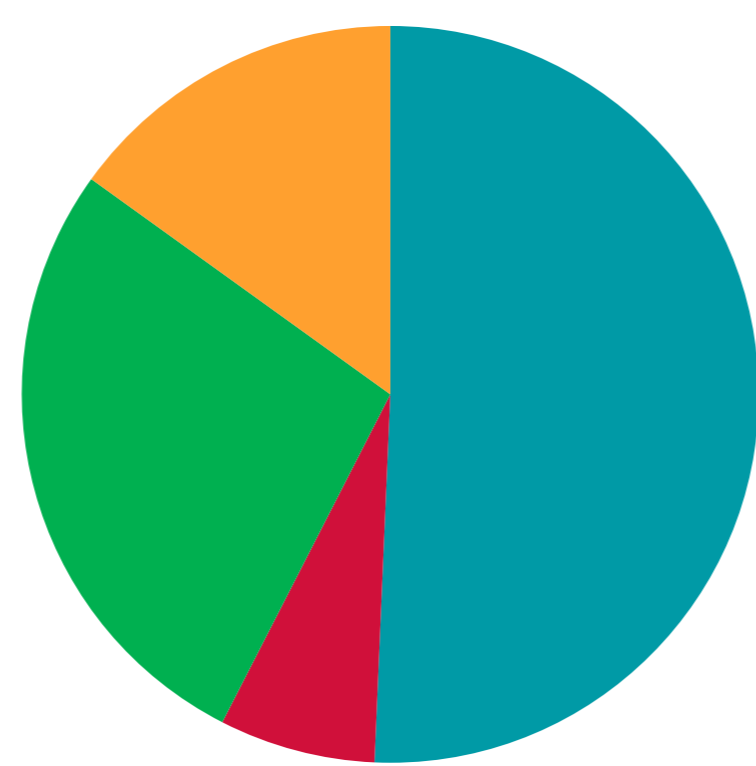
CONCLUSION

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

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

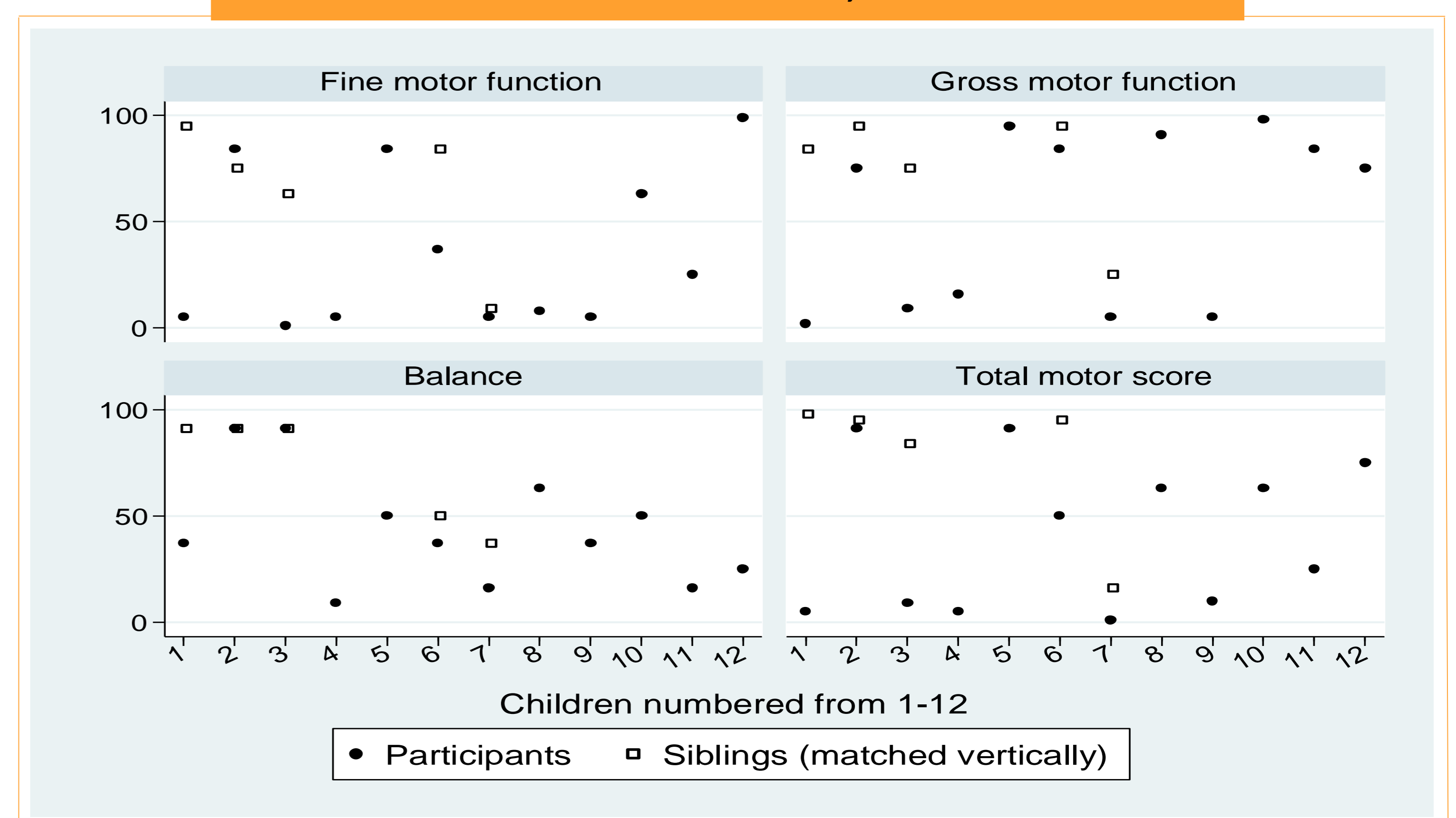


- Bayley Scale of Infant and Toddler Development, 3rd Edition (age 0-2 years = 37)
- DP-3 Developmental Profile (age >2 years =20)
- Wechsler's Intelligent Scale for Children, 4rd edition (age ≥6 N= 5)
- Movement ABC to assess motor function (age 3-17 years, n=10).

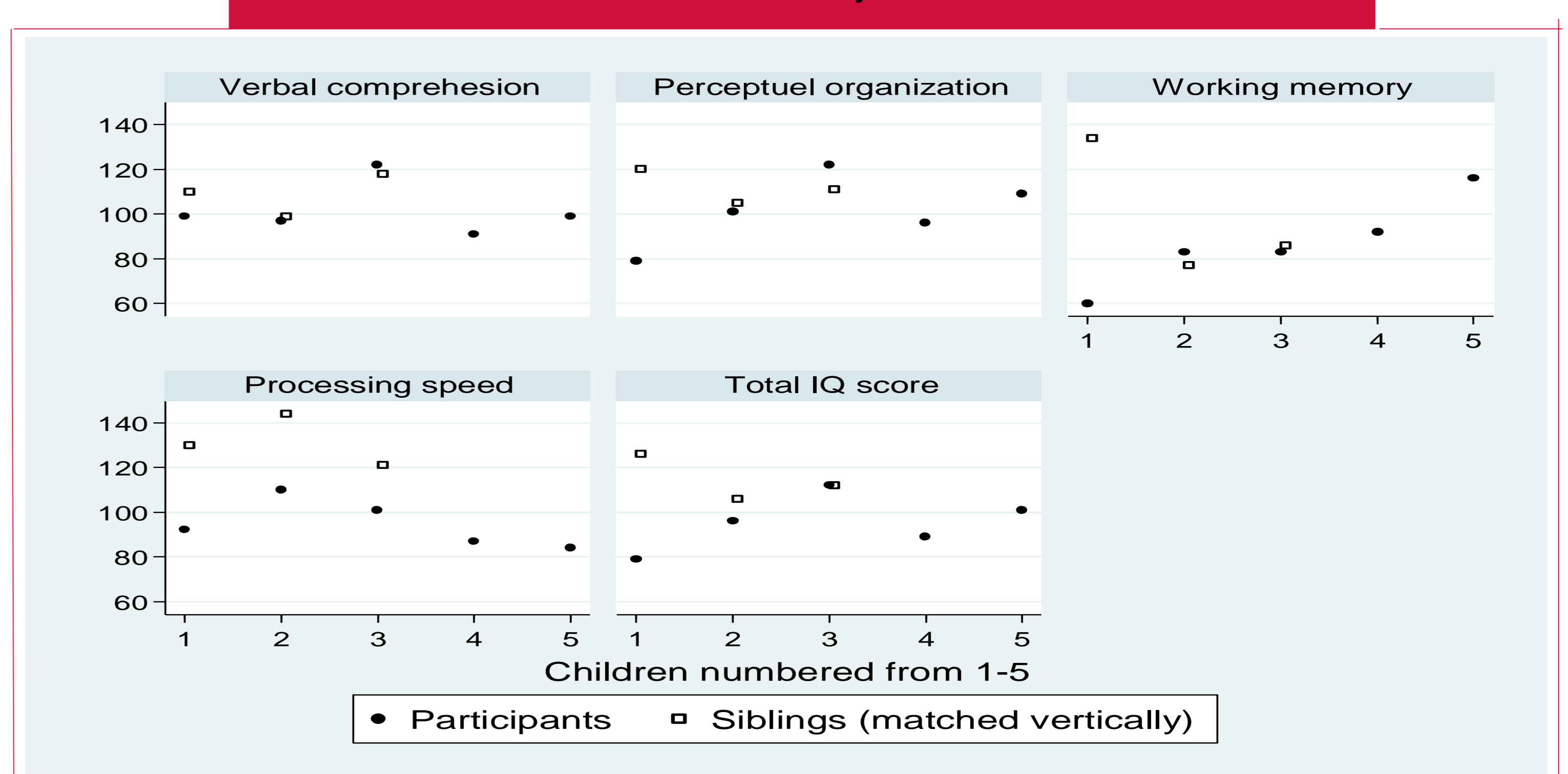
Normal development ** = IQ > 90, Bayley > 40 percentile or M-ABC > 40 percentile or DP3 > 85
Moderate disabled*** =IQ: 80 – 90 Bayley or M-ABC: 15 - 40 percentile or DP3: 60 – 85.
Severe disabled ***IQ < 80 or Bayley or M-ABC < 15 percentile or DP 3 < 60.

RESULTS cont.

Movement ABC-2, DK-SW



WISC-IV, DK



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

DP 3: delayed < 70, below average 70-84, average 85-115, above average 116-130, well above average >130.

Overall outcome for children with CHI.

	Total N= 67	Nordic N= 12	Foreign in DK N=22	Russia N= 32	Early onset N=42	Late onset N=25	K-ATP N=31	Other genetics N=36
Normal development	22(33)	7(58)	5(23)	10(31)	27(64)	13(52)	22(71)	18(50)
Moderate disabled	5(7)	0(0)	0(0)	5(16)	1(3)	4(16)	1(3)	4(11)
Severe disabled	40(60)	5(42)	17(77)	17(53)	14(33)	8(32)	8(26)	14(39)

Disclose statement: We have nothing to declare.

