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Arch Dis Child Fetal Neonatal Ed. Author manuscript; available in PMC 2015 September 15.

Published in final edited form as:

Arch Dis Child Fetal Neonatal Ed. 2014 January ; 99(1): F54–F58. doi:10.1136/archdischild-2013-304054.

Long-term developmental outcome of children with a fetal diagnosis of isolated inferior vermian hypoplasia

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Abstract

Objectives—Isolated inferior vermian hypoplasia (iiVH) is one of the most common fetal cerebellar anomalies presenting for fetal neurological counselling with controversial postnatal neurodevelopmental outcome. In the present study, we characterised the long-term neurodevelopmental outcome of prenatally diagnosed iiVH at school age.

Design and patients—We prospectively followed 20 children with fetal MRI diagnosis of iiVH including their postnatal MRI result and developmental outcome at school age (mean 6.1 years±1.9 years SD) using a comprehensive age-appropriate developmental testing battery, which encompassed cognitive, language, social and behavioural domains. Parental stress level and socioeconomic status were also evaluated.

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Contributors TT: Medical record review, MRI review, data collection, data/statistical analysis, manuscript preparation/revision. CL: Recruitment, developmental testing, medical record review, data collection, data/statistical analysis, manuscript preparation/revision. NRS: Developmental testing, data analysis, manuscript preparation/revision. RLR: MRI review, manuscript preparation/revision. AJdP: Recruitment, medical record review, MRI review, data/statistical analysis, manuscript preparation/revision

Competing interests None.

Ethics approval IRB at Boston Children's Hospital, Harvard Medical School.

Provenance and peer review Not commissioned; externally peer reviewed.

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Results—All children with postnatally confirmed iiVH had a normal neurodevelopmental outcome. A subgroup of children (2/20) who demonstrated cognitive delays and behavioural impairments had more extensive cerebellar malformation. Despite a normal developmental outcome, the parents of children with postnatally confirmed iiVH had higher parental stress compared with those parents whose children had normal postnatal MRI.

Conclusions—Children with postnatally confirmed iiVH show age appropriate functioning at school age. Postnatal MRI is important to confirm the diagnosis of iiVH and to exclude associated anomalies that impact neurodevelopmental outcome. A diagnosis of iiVH is associated with persistent elevated parental stress despite normal developmental outcomes in these children suggesting the need for ongoing parental support.

INTRODUCTION

Isolated inferior vermian hypoplasia (iiVH) is one of the most common fetal cerebellar anomalies detected by obstetrical ultrasound and presenting for prenatal counselling.[1–3] Despite its prevalence, the long-term neurodevelopmental consequences of iiVH remain poorly defined and controversial.[4,5] This is in part due to the difficulties in reliably diagnosing iiVH, especially by antenatal ultrasound, which provides relatively poor visibility of the posterior fossa structures.[6] Moreover, the MRI diagnosis of iiVH has a high false-positive rate.[4] Even at autopsy, confirmation of this diagnosis may be difficult given the challenge of preserving posterior fossa structures at fetopsy.[7] Additionally, inconsistencies in the classification of cerebellar anomalies further obscure the picture.[1,2,4,5,8,9] Finally, few studies have examined the long-term outcome of children diagnosed in utero with iiVH.

In a previous study, we examined the neuroimaging and neurodevelopmental outcomes of a cohort of infants (mean age of 19.8 months) with a prenatal diagnosis of iiVH.[4] We reported that a prenatal diagnosis of iiVH using second trimester MRI may overdiagnose the condition, with a false-positive rate of 32%, supported the recommendation for follow-up postnatal MRI. Although children with postnatally confirmed iiVH had overall lower mean developmental performance compared with infants with normal postnatal MRI, all children were free of major neurodevelopmental impairment and disability suggesting a relatively benign outcome at preschool age.[4] However, the extent to which these preschool developmental outcomes are transient or persist at school age remains unclear. We therefore sought to characterise the long-term neurodevelopmental outcome of iiVH by using standardised age appropriate assessments in our earlier cohort of infants with iiVH at school age.

METHODS AND PATIENTS

Selection criteria and procedures

We studied a total of 20 children with fetal MRI diagnosis of iiVH referred to Boston Children's Hospital. This included longitudinal follow-up of 17/19 children from our original cohort[4] (two were lost to follow-up) and an additional three fetuses who were subsequently diagnosed with iiVH and recruited in our study (overall 90.9% follow-up rate).

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All cases referred to our institution had fetal MRI. Using fetal MRI, iiVH was diagnosed when there was partial absence of the inferior cerebellar vermis without any apparent anomalies in cerebellar hemispheres, posterior fossa cystic lesions, supratentorial anomalies or other systemic malformations. We assessed the midline sagittal view vermian development from the caudal extent of the inferior vermis over the 4th ventricle, which findings were also confirmed on axial and coronal imaging.[4] All prenatal and postnatal MRI studies were reviewed by a paediatric neuroradiologist (RLR). We contacted the families and sought written informed consent for follow-up developmental evaluations. The study was approved by the IRB.

Medical record review

Medical record reviews and medical history questionnaires were conducted to obtain pertinent clinical information (eg, referral diagnosis, gender, birth weight, presence of visual and hearing abnormalities, any significant medical problems and the need for special education services).[4]

Developmental evaluations

Developmental outcome measures comprised the battery of standardised instruments described below. The examiners were blinded to past medical history and to the fetal-neonatal imaging findings. All children underwent either the Wechsler Preschool and Primary Scale of Intelligence—Third Edition (WPPSI-III) or Wechsler Intelligence Scale for Children—Fourth Edition (WISC-IV)[10][11] depending on the child's age (WPPSI-III for children between 2 years 6 months and 5 years 11 months; WISC-IV for children >6 years). We calculated Full Scale IQ (FS-IQ), General Language Composite or Verbal IQ (V-IQ), Performance IQ (P-IQ) and Processing Speed Quotient.[10] A score of <2SD of the normative mean is defined as abnormal. The WISC-IV additionally offers a Working Memory Index.

The Vineland Adaptive Behaviour Scale-II (VABS-II) is a discriminative norm-referenced measure of functional status in communication, daily living, socialisation and motor skills in children 0–18 years of age.[12] A score of <1.5SD below the normative mean (<78) is defined as abnormal.

The Behavior Rating Inventory of Executive Function (BRIEF) is a parental questionnaire used to determine if a child has a learning disability, attention deficit, memory impairment or some combination of these problems.[13] The cut-off is set ≥64 as clinically significant.[13]

The Child Behavior Checklist is a parental report of a child's behavioural problems. It offers externalising and internalising problem behaviour scores.[14] The clinical range is defined as T scores of 64, the borderline range as T scores from 60 to 63 and the reference range as T scores of <60.

The Social Communication Questionnaire is a parent-report screening measure for autism spectrum disorders.[15][16] A cut-off score of <15 is abnormal for this screening.[16]

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The Parental Stress Index (PSI) is a parent-report screening and diagnostic assessment measure of the relative magnitude of stress in the parent–child relationship.[17] Scores >85 is considered to be indicative of elevated levels of stress.

Socioeconomic status

The two-factor Hollingshead Index of Social Status was used to determine socioeconomical status (SES).[18] The index scores can range from 11 to 77 with lower scores representing higher SES.

Statistical analysis

Statistical analysis was performed using statistical software SAS V.9.2. Descriptive statistics were used to characterise the study cohort. Associations of two variables were tested using the Fisher's exact test, the two-sample t test or the Wilcoxon signed-rank test depending on the nature of variables. p Value<0.05 was considered significant.

RESULTS

Characteristics of the cohort

We performed follow-up developmental testing in 20 children at a mean age of 6.1 years (±1.9 years SD); 13 were male (65%). The median gestational age (GA) at birth was 39.5 weeks (range 36.5–41.0 weeks) and mean birth weight was 3481 g (±452 g SD). The prenatal diagnosis of iiVH was made at a median GA of 20.4 weeks (range 18.4–26.0 weeks). Six children (30%) had normal postnatal MRI studies. There was no difference in GA at diagnosis between children with con-firmed iiVH and those with normal postnatal MRI studies (20.9±0.5 vs 20.8±0.7 weeks, mean±SD, p=0.91). Of the 14 with abnormal postnatal MRI, 12 had a confirmed postnatal diagnosis of iiVH, while two children (10%) were found to have additional brain abnormalities on postnatal MRI that were not present on the prenatal MRI. Both children presented with bilateral dysplastic (small and poorly foliated) cerebellar hemispheres and mild pontine hypoplasia.

Neurodevelopmental outcome of children with a fetal diagnosis of iiVH

The neurodevelopmental and behavioural outcomes are summarised in tables 1 and 2. Overall, the majority of children (17/20 subjects) had normal developmental and behavioural scores, and were attending a regular preschool or primary school. Neither parental education nor occupation mediated outcomes.

Of the 12 subjects with postnatally confirmed iiVH, six were male and six were female. No gender difference was seen in neurodevelopmental testing.

Three children (15%) demonstrated abnormal neurodevelopment summarised in table 3. Two of them had more extensive cerebellar hemispheric malformations. One was a female with a chromosomal anomaly (microdeletion and microtrisomy in chromosome 10) (table 3. case 1) and another was a male (case 2). Both demonstrated greater behavioural dysfunction without apparent cognitive impairments. The third child was a male and had normal postnatal MRI (case 3). He was diagnosed with autism and had significant cognitive, social–

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behavioural impairment and received Applied Behavioral Analysis therapy. Parental stress was elevated in all three families.

Relationship between postnatal MRI findings and neurodevelopmental outcome

We compared the neurodevelopmental outcome in the subgroup of children with confirmed iiVH (n=12) and those children with normal postnatal MRI studies (n=5) (table 4). We excluded the one child with autism and the two children with additional cerebellar hemispheric anomalies detected on postnatal MRI, as this is likely to be an important confounder. Baseline and demographic characteristics were not significantly different between the two groups including age at fetal MRI diagnosis, mean GA at birth (39.3 vs 39.0 weeks), birth weight (3560 g vs 3363 g) and age at testing (6.6 vs 5.6 years of age). There was also no difference in neurodevelopmental outcome and parental social status. Moreover, there was higher parental stress for parents of children who had a postnatal MRI diagnosis of confirmed isolated iiVH compared with those parents whose children had a normal postnatal MRI (71.0 vs 45.2, p=0.001).

DISCUSSION

In this study, we describe a favourable neurodevelopmental outcome among children with iiVH as they enter school age. We used an age appropriate battery of standardised tests to confirm the stability of our findings in this well characterised population in early childhood, that is, before they were amenable to this level of testing. To the best of our knowledge, this is the first prospective study delineating the longitudinal neurodevelopmental outcome of children with iiVH.

Previous studies report controversial long-term neurodevelopmental outcome of iiVH, as some studies have suggested a benign outcome[3519–22] while others have reported poor outcome.[2324] Using a prospective study design and comprehensive standardised outcome measures, we demonstrate age-appropriate developmental outcomes among fetuses diagnosed with iiVH and confirmed postnatally. A small subgroup of our subjects (2/20) with the fetal diagnosis of iiVH had an abnormal neurodevelopmental outcome; however, this was associated with additional cerebellar hemispheric dysplasia and pontine hypoplasia detected on postnatal MRI studies. Such subtle findings may be missed by fetal MRI due to the small size of the structure and the exuberant ongoing cerebellar developmental processes during the third trimester (described below). This further underscores the need for confirmatory postnatal imaging.

Protracted fetal and postnatal cerebellar development

The cerebellum has a complex and protracted development that may contribute to the relatively high false-positive rate previously reported for the fetal diagnosis of iiVH[4] and overlooked associated malformations. Cerebellum has protracted morphological development that continues throughout the fetal period and into the early postnatal years. A retrospective ultrasound study has suggested that rostral–caudal development of the vermis is completed by 18 weeks[25]; however, this observation has been challenged recently by fetal MRI study.[26] Interestingly, several postnatal studies have reported ‘normalisation’ of

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prenatally diagnosed iiVH,[42127] suggesting that vermian growth may persist beyond 18 weeks. Because the lowest GA at diagnosis is 18.4 weeks in our study cohort, vermian development may be still in process. Lack of knowledge when that vermian growth completes might necessitate the recommendation of later gestation imaging, if situation allows. This is an intriguing question awaiting further study.

Elevated persistent parental stress

There was a persistent high level of parental stress (7/20, 35%) at a mean follow-up age of 6 years, and even when the neurodevelopmental outcome was benign. Not surprisingly, elevated parental stress was especially prevalent in cases where postnatal MRI confirmed vermian or cerebellar abnormality. Prenatal diagnosis of a congenital malformation is known to associate with higher acute and chronic parental psychological distress,[28] which might be worsened by unemployment and lower education level.[29] In our study, even the parents of children with a normal neurodevelopmental outcome in the years following the diagnosis suffered from higher stress levels regardless of their socioeconomic status. This suggests that the diagnosis of iiVH itself carries a significant and long-lasting emotional burden on these families despite the fact that their children are developing normally. Ongoing support and anticipatory guidance should be provided for these families prenatally and postnatally.

Limitations

Although this is the first prospective study to describe a favourable school-age outcome for children diagnosed prenatally with iiVH, given our small sample size, follow-up studies with larger cohorts are needed to confirm these observations. Our overall sample size was small especially when considering the size of our subgroups (14 normal and six abnormal postnatal scans). We diagnosed iiVH using qualitative, conventional assessment of the posterior fossa on fetal MRI that may have overdiagnosed iiVH. On the contrary, our findings are applicable to most of the clinical practices using non-quantitative fetal imaging. Future studies using advanced cerebellar MRI analyses warrant further investigation.

CONCLUSION

This study shows that children with postnatal confirmation of iiVH have normal cognitive, language, social and behavioural outcomes. This study also highlights the importance of postnatal MRI and karyotyping to confirm the diagnosis of iiVH. Additional, more extensive cerebellar malformations or chromosomal anomalies were strongly associated with worse neurodevelopmental outcome. Finally, despite the benign functional outcome of children with iiVH, the parents of these children carry elevated and enduring burden of stress. This finding emphasises the importance of pursuing improved fetal diagnosis and prognostication, as well as the need for ongoing parental support and anticipatory guidance.

Acknowledgments

Funding This study was supported by a grant from National Institute of Health (NINDS 1K24NS057568-01 (AJD)). Additional funding was provided by American Academy of Neurology (Clinical Research Training Fellowship, TT), Baby Alex Foundation (TT) and William Randolph Hearst Fund (TT).

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REFERENCES

1. Patel S, Barkovich AJ. Analysis and classification of cerebellar malformations. AJNR Am J Neuroradiol. 2002; 23:1074–87. [PubMed: 12169461]
2. Alkan O, Kizilkilic O, Yildirim T. Malformations of the midbrain and hindbrain: a retrospective study and review of the literature. Cerebellum. 2009; 8:355–65. [PubMed: 19337779]
3. Bolduc ME, Du Plessis AJ, Sullivan N, et al. Spectrum of neurodevelopmental disabilities in children with cerebellar malformations. Dev Med Child Neurol. 2011; 53:409–16. [PubMed: 21418200]
4. Limperopoulos C, Robertson RL, Estroff JA, et al. Diagnosis of inferior vermian hypoplasia by fetal magnetic resonance imaging: potential pitfalls and neurodevelopmental outcome. Am J Obstet Gynecol. 2006; 194:1070–6. [PubMed: 16580298]
5. Bolduc ME, Limperopoulos C. Neurodevelopmental outcomes in children with cerebellar malformations: a systematic review. Dev Med Child Neurol. 2009; 51:256–67. [PubMed: 19191827]
6. Adamsbaum C, Moutard ML, Andre C, et al. MRI of the fetal posterior fossa. Pediatr Radiol. 2005; 35:124–40. [PubMed: 15565345]
7. Griffiths PD, Variend D, Evans M, et al. Postmortem MR imaging of the fetal and stillborn central nervous system. AJNR Am J Neuroradiol. 2003; 24:22–7. [PubMed: 12533322]
8. Barkovich AJ, Kjos BO, Norman D, et al. Revised classification of posterior fossa cysts and cystlike malformations based on the results of multiplanar MR imaging. AJR Am J Roentgenol. 1989; 153:1289–300. [PubMed: 2816648]
9. Parisi MA, Dobyns WB. Human malformations of the midbrain and hindbrain: review and proposed classification scheme. Mol Genet Metab. 2003; 80:36–53. [PubMed: 14567956]
10. Lichtenberger EO. General measures of cognition for the preschool child. Ment Retard Dev Disabil Res Rev. 2005; 11:197–208. [PubMed: 16161087]
11. Wechsler, D. Manual for the Wechsler Intelligence Scale for children—revised. Psychological Corporation; New York: 1974.
12. Msall ME. Measuring functional skills in preschool children at risk for neurodevelopmental disabilities. Ment Retard Dev Disabil Res Rev. 2005; 11:263–73. [PubMed: 16161097]
13. Gioia GA, Isquith PK, Guy SC, et al. Behavior rating inventory of executive function. Child Neuropsychol. 2000; 6:235–8. [PubMed: 11419452]
14. Chandler S, Charman T, Baird G, et al. Validation of the social communication questionnaire in a population cohort of children with autism spectrum disorders. J Am Acad Child Adolesc Psychiatry. 2007; 46:1324–32. [PubMed: 17885574]
15. Berument SK, Rutter M, Lord C, et al. Autism screening questionnaire: diagnostic validity. Br J Psychiatry. 1999; 175:444–51. [PubMed: 10789276]
16. Norris M, Lecavalier L. Screening accuracy of Level 2 autism spectrum disorder rating scales. A review of selected instruments. Autism. 2010; 14:263–84. [PubMed: 20591956]
17. Loyd BH, Abidin RR. Revision of the parenting stress index. J Pediatr Psychol. 1985; 10:169–77. [PubMed: 4020601]
18. Schneider JA. Rewriting the SES: demographic patterns and divorcing families. Soc Sci Med. 1986; 23:211–22. [PubMed: 3749974]
19. Estroff JA, Scott MR, Benacerraf BR. Dandy-Walker variant: prenatal sonographic features and clinical outcome. Radiology. 1992; 185:755–8. [PubMed: 1438757]
20. Keogan MT, DeAtkine AB, Hertzberg BS. Cerebellar vermian defects: antenatal sonographic appearance and clinical significance. J Ultrasound Med. 1994; 13:607–11. [PubMed: 7933028]
21. Ecker JL, Shipp TD, Bromley B, et al. The sonographic diagnosis of Dandy-Walker and Dandy-Walker variant: associated findings and outcomes. Prenat Diagn. 2000; 20:328–32. [PubMed: 10740206]
22. Sasaki-Adams D, Elbabaa SK, Jewells V, et al. The Dandy-Walker variant: a case series of 24 pediatric patients and evaluation of associated anomalies, incidence of hydrocephalus, and developmental outcomes. J Neurosurg Pediatr. 2008; 2:194–9. [PubMed: 18759601]

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23. Klein O, Pierre-Kahn A, Boddaert N, et al. Dandy-Walker malformation: prenatal diagnosis and prognosis. Childs Nerv Syst. 2003; 19:484–9. [PubMed: 12879343]
24. Has R, Ermis H, Yuksel A, et al. Dandy-Walker malformation: a review of 78 cases diagnosed by prenatal sonography. Fetal Diagn Ther. 2004; 19:342–7. [PubMed: 15192294]
25. Bromley B, Nadel AS, Pauker S, et al. Closure of the cerebellar vermis: evaluation with second trimester US. Radiology. 1994; 193:761–3. [PubMed: 7972820]
26. Babcook CJ, Chong BW, Salamat MS, et al. Sonographic anatomy of the developing cerebellum: normal embryology can resemble pathology. AJR Am J Roentgenol. 1996; 166:427–33. [PubMed: 8553961]
27. Carroll SG, Porter H, Abdel-Fattah S, et al. Correlation of prenatal ultrasound diagnosis and pathologic findings in fetal brain abnormalities. Ultrasound Obstet Gynecol. 2000; 16:149–53. [PubMed: 11117085]
28. Skari H, Malt UF, Bjornland K, et al. Prenatal diagnosis of congenital malformations and parental psychological distress—a prospective longitudinal cohort study. Prenat Diagn. 2006; 26:1001–9. [PubMed: 16958144]
29. Skreden M, Skari H, Malt UF, et al. Long-term parental psychological distress among parents of children with a malformation–a prospective longitudinal study. Am J Med Genet A. 2010; 152A: 2193–202. [PubMed: 20803642]

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What is known about this topic

▶ Isolated inferior vermian hypoplasia is one of the most common fetal cerebellar anomalies diagnosed by fetal MRI, but its long-term neurodevelopmental prognosis is significantly controversial.

What this study adds

▶ Children with postnatally confirmed isolated inferior vermian hypoplasia have favourable neurodevelopmental function at school age.

▶ Postnatal MRI is important to confirm the diagnosis of inferior vermian hypoplasia and presence of associated anomalies that impact neurodevelopmental outcome.

▶ A diagnosis of isolated inferior vermian hypoplasia is associated with persistent elevated parental stress despite normal development outcomes in these children.

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Table 1

Cognitive outcomes of children diagnosed with fetal isolated inferior vermian hypoplasia (N=20)

Domains in test	WPPSI (Mean±SD)	WISC (Mean±SD)	Abnormal (cut-off <75, 1.5SD)
Verbal IQ	104.0±15.9	115.1±12.5	1/20 (5.0%)
Performance IQ (non-verbal)	100.4±19.7	100.4±29.8	0/20 (0.0%)
Processing speed quotient	98.0±13.5	107.9±13.1	1/20 (5.0%)
Full scale IQ	104.2±14.6	114.0±12.5	1/20 (5.0%)
WISC working memory index	N/A (WISC only)	109.9±12.5	0/20 (0.0%)

WISC, Wechsler Intelligence Scale for Children—4th edition; WPPSI, Wechsler Preschool and Primary Scale of Intelligence—3rd edition.

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Table 2

Functional and behavioural outcomes of children with fetal diagnosis of isolated inferior vermian hypoplasia (N=20)

Tests	(Mean±SD)	Abnormal (cut-off <78, 1.5SD)
Vineland Adaptive Behavior Scales-II		
Communication	97.2±11.4	2/20 (10.0%)
Daily living skills	93.3±7.5	1/20 (5.0%)
Socialisation	95.2±10.8	1/20 (5.0%)
Motor	95.8±8.3	1/20 (5.0%)
Working memory	93.9±7.3	1/20 (5.0%)
Child Behavior Check List		
Internalising behavioural problems	46.1±10.6	Abnormal 1/20(5.0%)
Externalising behavioural problems	45.1±8.3	Abnormal 0/20 (5.0%)
Total	45.9±9.8	Abnormal 2/20 (10.0%)
	Median, (1st, 3rd quartile)	
Social communication questionnaire	3.00 (1.25, 4.00)	Abnormal 1/20(5.0%)
BRIEF global executive composite	44 (37, 60)	>65 4/20 (20.0%)
Index of social status		
Mother's education	18.0 (15.0, 21.0)	
Mother's occupation	40.0 (40.0, 40.0)	
Father's education	19.4 (15.8, 21.0)	
Father's occupation	35 (30.0, 45.0)	
Parental Stress Index	67.5 (46.5, 86.0)	>85 (High stress) 7/20 (35.0%)

BRIEF, Behavior Rating Inventory of Executive Function.

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Table 3

Neuropsychological outcomes in the subgroup of children with more extensive cerebellar dysgenesis (n=2) and one child with normal postnatal MRI (n=1)

Case	1	2	3
Prenatal MRI	Isolated iVH	Isolated iVH	Isoalated iVH
Postnatal MRI	iVH+cerebellar hemisphere anomalies	iVH+cerebellar hemisphere anomalies	normal
Gender	Female	Male	Male
Note	del distal end of 10q, trisomy at 10q26.1		autism
School setting	Special education	Special education	Special education with ABA
WPPSI			
Composite scores	84	85	64*
Performance IQ (non-verbal)	87	89	90
Processing speed quotient	82	84	68*
Full scale IQ	85	87	75
Vineland Adaptive Behavior Scales-II			
Communication	76	87	76
Daily living skills	75	77	83
Socialisation	87	89	59*
Motor	75	84	107
Adaptation	77	90	81
Child Behavior Check List			
Internalising behavioural problems	55	61	66*
Externalising behavioural problems	55	55	46
Total	58	61	60
Social communication questionnaire	5	3	18*
BRIEF global executive composite	75	60	80
Parental Stress Index	86*	89*	105*

ABA, applied behavioral analysis; BRIEF, behavior rating inventory of executive function; iVH, inferior vermian hypoplasia; WPPSI, Wechsler Preschool and Primary Scale of Intelligence—3rd edition.

*Demotes abnormal neurodevelopmental outcome results.

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Table 4

Comparison of neuropsychological assessment in children with postnatally confirmed iiVH (n=12) and those with normal postnatal MRI (n=5) in postnatal MRI

	iiVH (n=12) (Mean±SD)	Normal (n=5) (Mean±SD)	p Value
WPPSI & WISC			
Composite scores	108.1±13.0	110.0±7.7	0.64
Performance IQ (non-verbal)	102.3±24.3	102.8±9.4	0.53
Processing speed quotient	100.8±10.6	103.0±12.7	0.63
Full scale IQ	107.9±12.9	108.2±10.8	0.52
WISC Working Memory Index (iiVH n=5, normal n=2)	107.4±13.8	116.0±8.5	0.80
Vineland Adaptive Behavior Scales-II			
Communication	101.0±10.2	98.6±6.0	0.28
Daily living skills	96.7±4.0	94.2±3.6	0.12
Socialisation	98.3±6.4	97.6±6.8	0.42
Motor	96.3±6.3	100.2±6.4	0.24
Adaptation	95.8±6.2	96.0±3.7	0.54
Child Behavior Check List			
Internalising behavioural problems	43.1±3.0	48.2±4.2	0.40
Externalising behavioural problems	44.7±2.4	42.7±3.4	0.57
Total	43.8±2.7	45.5±3.8	0.73
Social communication questionnaire	2.5±1.7	2.2±1.3	0.35
	Median, (1st, 3rd quartile)	Median, (1st, 3rd quartile)	Wilcoxon rank test p Value
BRIEF global executive composite	44.5 (42.0, 57.5)	32 (30.0, 34.0)	0.002*
Parental Stress Index	73.0 (57.5, 86.0)	45 (42.5, 48)	0.007**

BRIEF, Behavior Rating Inventory of Executive Function; iiVH, isolated inferior vermian hypoplasia; WISC, Wechsler Intelligence Scale for Children—4th edition; WPPSI, Wechsler Preschool and Primary Scale of Intelligence—3rd edition.

* Significant ($p<0.05$), both group's scores were still within normal range.

** Significant ($p<0.05$).