Vineland Adaptive Behavior Scales to Identify Neurodevelopmental Outcomes in Children with Congenital Hypothyroidism

Original Article

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ABSTRACT

Background: Congenital hypothyroidism (CH) is the most common preventable causes of mental retardation in children. Screening and early treatment an important prognostic factors for normal neurodevelopment in children with CH. The Vineland Adaptive Behavior Scales Second Edition (VABS-II) is a parent report measure of adaptive functioning that could be used as a developmental screening tool in patients with CH. We have investigated the performance of VABS-II as a screening tool to identify developmental delay in children with CH. VABS-II questionnaires testing communication, daily living skills, social skills, motor skills and behaviour domains were completed by parents of 50 children with CH initially at time of diagnosis and follow up after six month with age distribution 3rd day of life - 12 months (CH, n = 35) and 13- 24 months (CH, n = 15). Both had positive results of screening program and positive values for thyroid function tests.

Objective: The aim of this study to assess and follow up neurodevelopmental abilities in children with congenital hypothyroidism using Vineland Adaptive Behavior Scales-Second Edition (Vineland-II).

Patients and Methods: We conducted a prospective study at Benha Children Hospital, Egypt. The study participants were 50 infants with CH and their mothers (patients group), and 50 age matched volunteers of healthy infants (control group). Data were collected to determine socioeconomic state, clinical picture at presentation, personal, family, obstetric history and assessment of neurodevelopment by using Vineland-II scale.

Results: About 64% of cases were permanent CH and about 14% of cases were transient CH. There was significant positive correlation between compliance score and the parameters of the physical and neurological development.

Conclusion: Our study adds further evidence to normalize TSH as soon as possible to avoid long-term neurodevelopmental outcomes.

Key Words: Congenital hypothyroidism, neurodevelopment, Vineland-II.

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INTRODUCTION

Congenital Hypothyroidism (CH) defined as dysfunction of the hypothalamic–pituitary–thyroid (HPT) axis present at birth, resulting in insufficient thyroid hormone production and with that, severe to mild thyroid hormone deficiency[1]. CH one of the most common preventable endocrinal causes of mental retardation[2].

The detection and treatment of newborns with hypothyroidism should be considered a pediatric emergency. If therapy is not begun soon after birth, developmental delay will result[3]. However, even in patients with CH who receive early treatment, subtle selective cognitive deficits and some signs of minimal brain damage have been reported in some studies[4].

Factors that influence the neurodevelopmental outcome in children with CH are the age at initiation of treatment, starting dose of levothyroxine, severity of disease, free T4 concentrations in first 2 years of life, and compliance to therapy[5].

The aim of this study to assess and follow up neurodevelopmental abilities in children with congenital

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hypothyroidism using Vineland Adaptive Behavior Scales-Second Edition (Vineland-II)[6].

PATIENTS AND METHODS

All diagnosed infants with congenital hypothyroidism age ranged from 3rd day to 2 years old. They presented for treatment and follow up at the Pediatric Endocrine Clinic Benha Children Hospital Al-Qalyopia, Egypt enrolled in the study as a patient group over a period of one year from September 2018 till September 2019.

The study patient group included 50 patients (26 male and 24 female) and age and sex matched volunteers of healthy infants included 50 participants (23 male and 27 female) were enrolled as a control group.

All the study participants were subjected to:

I. History and questionnaires: personal history and family history, clinical picture of congenital hypothyroidism at presentation, neurological examination

II. Assessment of the adaptive behavior: by using Vineland Adaptive Behavior Scales-Second Edition (Vineland-II).

RESULTS

Table 1: Comparison between initial Vineland-II and after 6 months in the congenital hypothyroidism patients.

Studied variable	Initial Vineland-ll		Vineland-ll after 6 months		Mc Nemar test	*P* value
	No.	%	No.	%		
Communication						
Receptive						
Adequate	48	96.0	50	100	2.04	0.500
Low moderate	2	4.00	0	0.00		
Expressive						
Adequate	19	38.0	35	70.0	10.3	0.001**
Low moderate	31	62.0	15	30.0		
Daily Living Skills						
Personal						
Adequate	42	84.0	49	98.0	5.98	0.016*
Low moderate	8	16.0	1	2.00		
Domestic						
Adequate	50	100	50	100	--	--
Low moderate	0	0.00	0	0.00		
Motor Skills						
Gross						
Adequate	31	62.0	37	74.0	1.65	0.198
Low moderate	19	38.0	13	26.0		
Fine						
Adequate	47	94.0	48	96.0	0.21	0.646
Low moderate	3	6.00	2	4.00		
Socialization						
Interpersonal Relationships						
Adequate	50	100	50	100	--	--
Low moderate	0	0.00	0	0.00		
Play and Leisure Time						
Adequate	18	36.0	26	52.0	2.60	0.096
Low moderate	32	64.0	24	48.0		
Adaptive Behavior Composite						
Adequate	31	62.0	41	82.0	8.12	0.017*
Low mild	6	12.0	0	0.00		
Low moderate	13	26.0	9	18.0		

*Significant **High significant

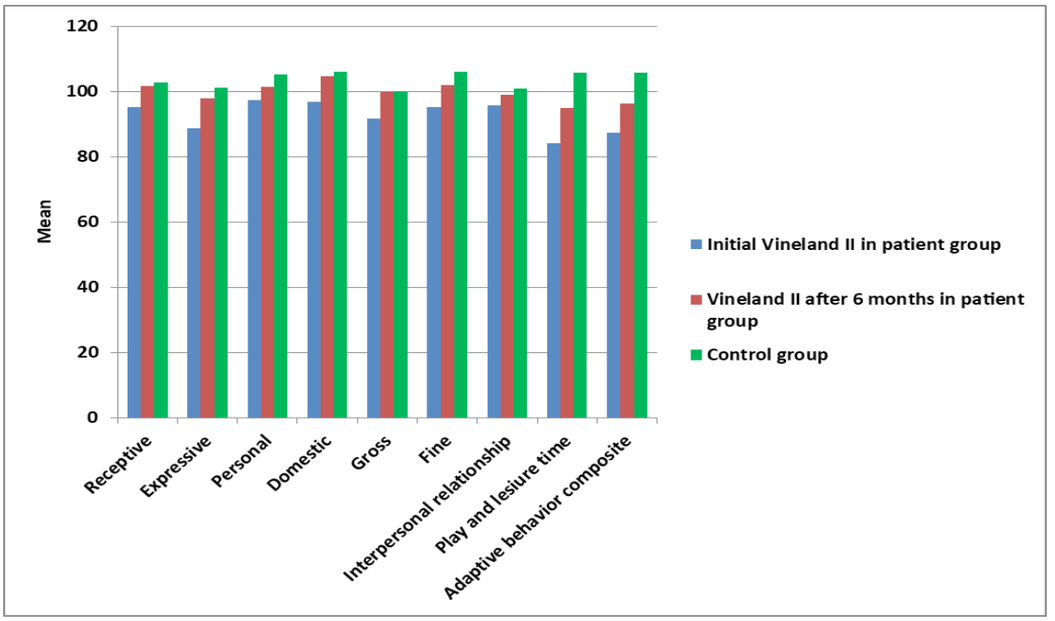

Fig. 1: Comparison between Vineland-ll of control group and Initial Vineland-II and after 6 months in the studied case group.

Table 2: Relation between initial Vineland-II and the Length of normalization period of TSH

Length of normalization period of TSH	Initial Vineland-ll						X2	*P* value
	Adequate		Low mild		Low moderate			
	No.	%	No.	%	No.	%		
Within 15 days	14	45.2	1	16.7	2	15.4		
16 – 30 days	10	32.3	1	16.7	8	61.5	9.52	0.049*
Above month	7	22.6	4	66.7	3	23.1		

*Statistical significance.

DISCUSSION

In our study we analyze early developmental milestones red flags, our results showed that 22% of cases with CH developed delay in language acquisition followed by 20% delay in Gross Motor Skills; mainly delay in both sitting without support (8%) and walking independently (8%), followed by 16% delay in Fine motor skills and 12% delay in social skills.

Results of our study showed that about 64% of cases have permanent CH and about 22% of cases were transient. The type of CH is not known yet in the remaining 14% of cases.

By reviewing the records in our study thyroid dysgenesis was founded in 74% of cases (agenesis in 44.4% and ectopic thyroid in 29.6% of cases) followed by dyshormonogenesis (16.8%) and antithyroid drug intake (11.1%) were the main causes of CH in. Farahat *et al.*[7] reported thyroid dysgenesis (agenesis in 52.2% and ectopic thyroid 28.4%) followed by dyshormonogenesis (11.94%) and antithyroid drug intake (7.46%) in Egypt.

In our study 64% of children with CH had no clinical manifestation while 36% of cases had clinical manifestation and the most common presenting symptoms before treatment were prolonged neonatal jaundice (44%) then enlarged tongue, umbilical hernia and constipation each of them represented with (11%). Most children with CH have little or no clinical manifestation of the disease at birth due to the trans- placental passage of maternal T4 and because most affected children have some functioning thyroid tissue.

We observed associated congenital malformation among 22% of CH cases and the most common congenital anomalies were cardiac anomalies 10% followed by hearing impairment 6%.

The criteria of CH severity in our study based on

levels of FT4 according to the new European Society for Pediatric Endocrinology and the European Society for Endocrinology consensus guidelines[1], most of the patients (46%) had mild form of CH while 38% of the patients were moderate form of CH and 16% were severe form of CH.

In our study, we used VABS-II for assessment of neurodevelopmental delay in children with CH in correlation to compliance. VABS-II individually correlated with developmental delay with high specificity, indicating accuracy of screening investigations for formal developmental assessments[8].

Vineland-II performed in our infants with CH, over six month period and correlated parent compliance and adherent to medication with objective clinical laboratory investigations. The Initial Vineland-ll scores in the studied patients at the first visit of assessment showed that most of the patients (62%) had adequate adaptive behavior composite while 26% were moderate low ¬1.0 -- 2.0- standard deviation from the mean and 12% of the patients were low mild deficit 2.0- or below standard deviation from the mean.

After six months of treatments Vineland-II repeated to studies patients show improving of Vineland- II scores as most of the patients (82%) became adequate in their adaptive behavior composite while 18% of the patients still in the moderate low adaptive category with no children below 2.0- standard deviation which represented as low mild deficit. Comparison between Initial Vineland- II and after 6 months in the studied patients showed improving of expressive subdomain of communication domain, personal sub-domain in Daily living Skills domain and Adaptive Behavior Composite significantly improved after six months. The study results were statistically significant for the relation between the initial Vineland-II and the length of normalization period of TSh as shown in table (2), therefore our study emphasizes further evidence of the importance of normalizing the TSH as soon as possible to avoid any long-term neurodevelopmental insults.

There were no statistical differences in Communication and Motor Skills domains. On the other hand, scores in Daily Living Skills, Socialization and Adaptive Behaviour Composite, in children with CH were significantly lower than in normal population ($P < 0.05$) as reported by Almedia[9].

Azar-Kolakez *et al.*[10] reported that adequately treated infants with CH grow and develop normally. The psychometric outcome is much improved over the prescreening era, but some severely affected infants or those who are inadequately treated or those with poor compliance to treatment especially in the first two or three years of life have IQs below those of normal children. Similar results were detected by Dimitropoulos *et al.* in Switzerland[11].

Gruters and Krude[12] reported that the outcome of the affected children with CH depends on optimal compliance of the parents and later in life of the patients themselves, another study from Brazil reported a clinically significant effect of the compliance to therapy on the final neurodevelopmental outcomes.

CONCLUSION

Vineland-II can be reliably used in neurodevelopmental follow up of CH patients to trigger formal developmental assessment. Our study adds further evidence to normalize TSH as soon as possible to avoid long-term neurodevelopmental outcomes.

CONFLICT OF INTEREST

There are no conflicts of interest.

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