

Behavioral assessment of children with Spondylo-epi-metaphyseal dysplasias using the child behavior checklist

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Abstract

SEMD is among the most important diseases that cause disproportionate short stature and varying degrees of skeletal deformities (Temtamy et al., 2007), These patients may also suffer from many associated behavioral problems. The effects of these complications on the patient's behavior in previous studies were not definitive.

Patients And Methods: This study is a descriptive and exploratory study involving children with SEMDs recruited from the Limb Malformations and Skeletal Dysplasia Clinic (LMSDC), Center of Excellence of Human Genetics, National Research Centre (NRC). The study included thirty seven cases. Patients were classified according to the disease severity and associated complications into three groups. The child behavior checklist DSM- Oriented Scales for children translated by Dr. Mona Elsheikh (Achenbach, 2001) was used to assist behavior problems of each child to determine the existence of any behavioral problems.

Results: The study included 37 cases of SEMDs, 7 cases of which had mild complications, 18 cases had moderate complications, and 12 cases had severe complications. The number of male patients was 22 and 15 were females. The study showed that these children suffer from several behavioral problems, especially the affective problems (62.1%).

Conclusion: This study emphasizes the importance of studying the behavioral problems associated with SEMDs which result from short stature and skeletal deformities. Early diagnosis is very important for referral of patients into specialists to begin prompt management in order to ease the associated behavioral problems.

Key Words: Skeletal dysplasia, SEMDs, behavior problems, child behavior checklist DSM- Oriented Scales for children.

التقييم السلوكي للأطفال الذين يعانون من مرض خلل العمود الفقري والنسيج الكردوسي والفرق كره وسى

باستخدام قائمة تقييم سلوك الطفل

المقدمة: تعتبر امراض خلل العمود الفقري والنسيج الكردوسي والفرق كره وسى SEMDs من بين اهم الامراض التى تسبب قصر غير متناسق فى القامة وتشوهات متعددة فى الهيكل العظمى بدرجات متفاوتة فى الشدة، وكذلك قد يعانى هؤلاء المرضى من العديد من المشكلات السلوكية المصاحبة. ولم يتم تحديد تأثير هذه المضاعفات على المرضى فى الدراسات السابقة بشكل قاطع.

الهدف: دراسة المشكلات السلوكية للأطفال المصابين بامراض SEMDs وتأثير شدة المضاعفات المختلفة للمرض عليهم.

طرق وادوات الدراسة: يشكل هذا البحث دراسة وصفية واستطلاعية شملت الأطفال الذين يعانون من SEMDs من المترددين على عيادة تشوهات الأطراف والعظم الوراثية بالمركز القومي للبحوث. وتضمنت الدراسة سبع وثلاثين حالة وقد تم تصنيفهم حسب شدة المضاعفات الى ثلاثة اقسام وتم عمل قائمة تقييم لفحص سلوك كل طفل وذلك لتحديد وجود اى مشلات سلوكية لديه.

النتائج: وقد تضمنت الدراسة عدد سبع وثلاثين حالة من امراض SEMDs منهم سبع حالات تعانى من مضاعفات بسيطة وثمان عشرة حالة تعانى من مضاعفات متوسطة واثنا عشرة حالة تعانى من مضاعفات شديدة. وكان عدد المرضى الذكور اثنان وعشرين حالة والانثى خمس عشرة حالة. وقد اظهرت الدراسة انه برغم اختلاف شدة المرض يعانى هؤلاء الأطفال من عدة مشكلات سلوكية وخاصة فى السلوك العاطفى مثل الاكتئاب الحاد والمزمن حيث وصلت نسبة المصابين او المعرضين للإصابة به الى ٦٢,١%.

الخلاصة: بينت هذه الدراسة أهمية دراسة المشكلات السلوكية المصاحبة لمرضى SEMDs الناتجة عن معاناتهم من قصر القامة والتشوهات العظمية المصاحبة. كما ان التشخيص المبكر مهم جدا لتحويل المرضى الى المختصين للبدء فى علاجهم ومساعدتهم.

Introduction:

Spondylo-epi-metaphyseal dysplasias (SEMDs) are clinically and genetically heterogeneous group of skeletal dysplasias characterized by defective growth and modeling of the spine and long bones. In SEMDs, disturbed growth can be recognized by abnormal radiographic findings within the epiphyses of long bones, the adjacent metaphyses and the vertebral bodies (Temtamy et al., 2007).

SEMDs represent a subgroup of osteochondrodysplasia, which is a heterogeneous group of conditions caused by impaired development of the osseous skeleton (Borochowitz et al., 2004).

Classification of SEMDs is nosologically difficult. By reviewing the Online Mendelian Inheritance in Man (OMIM) and London Medical Databases (LMD), SEMDs group comprises more than 80 distinct entities with differing modes of inheritance, all defined by the combination of vertebral, epiphyseal and or metaphyseal abnormalities (Omim 2018; LMD 2018).

The SEMDs often arise sporadically, but distinctive heritable forms with autosomal dominant, autosomal recessive or X-linked transmission have been reported. The severity varies among and within affected SEMD's kindred (Kennedy et al., 2005).

The presenting symptoms of SEMD patients are usually variable degrees of disproportionate short stature and skeletal deformities (Cormier-Daire, 2008).

The review of literature was inconclusive regarding whether the associated skeletal deformities and short stature in SEMDs were the reasons for the different behavior problems or not. Few studies were done with limitations to the more common types of skeletal dysplasias such as Achondroplasia or Osteogenesis imperfecta and their results were variable (Adelson, 2005).

Association of short stature due to skeletal dysplasia with psychological outcomes was studied by Wheeler et al (2003). The study found no evidence for increased rates of depression or anxiety in children with skeletal dysplasias.

No studies discussed the psychological effect of short stature and skeletal deformities which are commonly associated in patients with SEMDs till now.

Patients and Methods:

In this study, we checked the existence of behavioral changes in SEMDs patients and checked the correlation between the behavior changes and patients with SEMDs, who had variable degrees of short stature and skeletal deformities. Also, we tried to check the effect of severity of these SEMDs complications on the examined patient's behavior.

We studied 37 cases of SEMDs who were recruited from the Limb Malformations and Skeletal Dysplasia Clinic (LMSDC), Center of Excellence of Human Genetics, National Research Centre (NRC) from January 2015 to December 2017. Patients' age was between (6- 18) years and IQ was more than 70.

Patients were divided into mild, moderate and severe according to the severity of associated complications.

We also checked the degree of IQ of our patients, the effect of poor activities participation, social participation and poor school performance and the effect of sex difference between our studied cases in relation to their behavior problems to diagnose the assistant factors which may be associated.

We examined the patients using the child behavior checklist DSM-Oriented Scales for children translated by Dr. Mona Elsheikh (Achenbach, 2001) which is questionnaire assessing 6 subscales; affective problems (dysthymia and major depression), anxiety problems, somatic problems, attention deficit problems, oppositional defiant problems and conduct problems. Responses are recorded on a Likert scale: 0= Not True, 1= Somewhat or Sometimes True, 2= Very True or Often True. The standard scores are scaled so that 50 is average for the youth's age and gender, with a standard deviation of 10 points. Higher scores indicate greater problems.

For each syndrome, Internalizing (affective problems, anxiety problems and somatic problems) and Externalizing problem scales (attention deficit problems, oppositional defiant problems and conduct problems), and the total score, scores can be interpreted as falling in the normal (T score < 65), borderline (T score from 65 to 70), or clinical behavior (T score > 70). CBCL competence scale questionnaire assesses; activities, social and school performance is included as a part of CBCL of Achenbach (2001).

Statistical Analysis:

Qualitative data were statistically represented in terms number and percent. Comparison between difference groups in the presents study was done using Crosstabs with Chi-Square Test.

Correlation between various variables was done using spearman correlation coefficient (R). R: can take a range of values from 1 to -1. A value of 0 indicates that there is no association between the two variables. A value greater than 0 indicates a positive association. A value less than 0 indicates a negative association. 1 is a perfect positive and -1 is a perfect negative. Values between 0.7 and 1 (-0.7 and -1) indicates strong positive (negative). Values between 0.3 and 0.7 (-0.3 and -0.7) indicates moderate positive (negative). Values between 0.3 and 0 (-0.3 and 0) indicates weak positive (negative).

A probability value p value less than or equal to 0.05 was considered significant. All statistical calculations were done using computer program SPSS (Statistical Package for Social Science) statistical program version 16.0. Graphs were done using SPSS statistical program version 16.0 and Microsoft Excel program version 2016.

Results:

We studied 37 cases of SEMDs with variable degrees of skeletal deformities and short stature by the use of child behavior checklist DSM-Oriented Scales for children. Males were 22 (59.4%) cases and female were 15 cases (40.6%). Mild cases were 7 cases, moderate cases were 18 and severe cases were 12 cases.

Most of our patients showed more liability or clinical range to affective problems (62.1%) than other studied behavior problems. Anxiety problems were the next, followed by somatic, oppositional defiant and attention deficit, respectively. Two of our patients showed a liability to conduct problems.

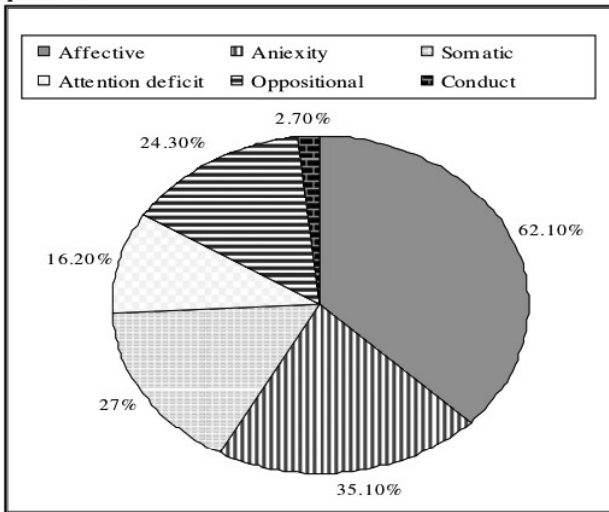


Figure (1) Pie chart illustrating the percentage of each behavioral problem in the studied SEMDs patients.

Table (1) CBCL DSM- Oriented Scale results:

CBCL DSM- Oriented Scales results:	Affective Problems	Anxiety Problems	Somatic Problems	Attention Deficit Score	Oppositional Defiant Problems	Conduct Problems
No of affected cases and its percentage:	23 cases (62.1%)	13 cases (35.1%)	10 Cases (27%)	6 cases (16.2%)	9 cases (24.3%)	2 cases (2.7%)
T Score Range:	50- 85	50- 80	50- 80	50- 75	50- 75	50- 70
Mean T scores and SD:	71.43±8.54	63.24±7.52	62.22±6.69	57.11±8.95	58.38±7.73	54.24±5.33
Ci:	15.23- 20.88	9.40- 14.27	9.90- 14.49	4.47- 8.64	5.52- 10.70	2.40- 6.05
P Value:	0.001	0.001	0.001	0.001	0.001	0.001
Mean T scores of internalizing, externalizing and total of all scales:	Mean T scores of internalizing scales, SD and p value			Mean T scores of externalizing scales, SD and p value		
	65.63±7.58 (p value= 0.000)			56.58±7.34 (p value= 0.001)		
Mean T scores of all scales, SD and P value:						
61.1±7.46 (p value= 0.001)						

ore of internalizing problems is considered borderline (65.63±7.58) and the mean T score of affective problems of SEMDs patients is considered clinically concerning compared to the general population (mean= 71.43± 8.54).

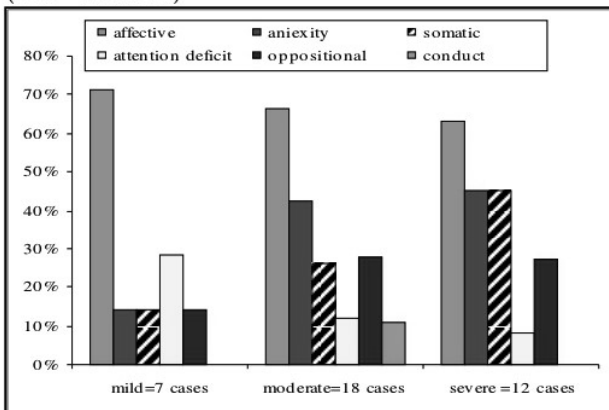


Figure (2) Bar chart illustrating each studied behavioral problem percentage in the three different groups of skeletal severity.

Table (2) Behavioral problems in relation to variable degrees of severity:

Behavioral Problems		Severity grade (N= 37)			R	P- Value
		Mild, 7 Cases	Moderate, 18 Cases	Severe, 12 Cases		
		No (%)	No (%)	No (%)		
Affective Problems	No	2 (28.6)	7 (38.9)	5 (41.7)	-0.85	0.653
	Yes	5 (71.4)	11 (61.1)	7 (58.3)		
Anxiety Problems	No	6 (85.7)	11 (61.1)	7 (58.3)	0.173	0.333
	Yes	1 (14.3)	7 (38.9)	5 (41.7)		
Somatic Problems	No	6 (85.7)	14 (77.8)	7 (58.3)	0.233	0.176
	Yes	1 (14.3)	4 (22.2)	5 (41.7)		
Attention Deficit	No	5 (71.4)	15 (83.3)	11 (91.7)	-0.187	0.334
	Yes	2 (28.6)	3 (16.7)	1 (8.3)		
Oppositional Defiant	No	6 (85.7)	13 (72.2)	9 (75)	0.064	0.775
	Yes	1 (14.3)	5 (27.8)	3 (25)		
Conduct Problems	No	7 (100)	16 (88.9)	12 (100)	-0.061	0.874
	Yes	0 (0)	2 (11.1)	0 (0)		

The comparison of the results of different behavioral problems in relation to SEMDs severity showed nearly no significant difference between mild, moderate and severe cases in the prevalence of behavioral problems.

The comparison of the results of different behavioral problems in relation to IQ of SEMDs patients were illustrated in figure (3) and table (3).

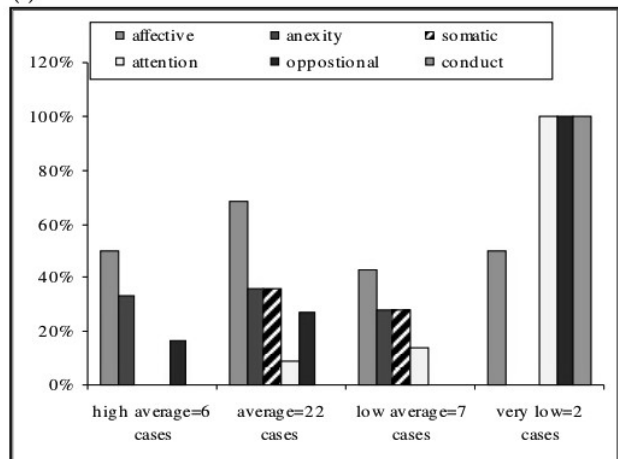


Figure (3) Bar chart illustrating behavioral problems percentage in relation to IQ grade.

Table (3) behavioral problems in relation to IQ grade:

Behavioral Problems		IQ grade (N= 37)				R	P Value
		High Average (IQ= 110- 119), 6 Cases	Average (IQ= 90- 109), 22 cases	Low Average (IQ= 80- 89), 7 Cases	Very Low (IQ= 70- 79), 2 Cases		
		No (%)	No (%)	No (%)	No (%)		
Affective Problems	Yes	3 (50.0)	15 (68.2)	3 (42.9)	1 (50.0)	0.073	0.617
	No	3 (50.0)	7 (31.8)	4 (57.1)	1 (50.0)		
Anxiety Problems	Yes	2 (33.3)	8 (36.4)	2 (28.6)	0 (0.0)	0.104	0.761
	No	4 (66.7)	14 (63.6)	5 (71.4)	2 (100.0)		
Somatic Problems	Yes	0 (0.0)	8 (36.4)	2 (28.6)	0 (0.0)	-0.090	0.268
	No	6 (100.0)	14 (63.6)	5 (71.4)	2 (100.0)		
Attention Deficit	Yes	0 (0.0)	2 (9.1)	1 (14.3)	2 (100.0)	-0.377	0.003
	No	6 (100.0)	20 (90.9)	6 (85.7)	0 (0.0)		
Oppositional Defiant Problems	Yes	1 (16.7)	6 (27.3)	0 (0.0)	2 (100.0)	-0.070	0.033
	No	5 (83.3)	16 (72.7)	7 (100.0)	0 (0.0)		
Conduct Problems	Yes	0 (0.0)	2 (9.1)	0 (0.0)	2 (100.0)	-0.296	0.001
	No	6 (100.0)	20 (90.9)	7 (100.0)	0 (0.0)		

P values were significantly correlated with the attention deficit

problems, oppositional defiant problems and conduct problems in relation to IQ degrees.

Activities, social and school performance were assessed by the use of the child behavior checklist Competence Scales (Figure (4)& Table (4), (6) and (7) results showed that:

- ✎ 31 cases (83.7%) had poor activities participation: 20 cases (64.5%) were males and 11 females (35.5%)
- ✎ 19 cases (51.3%) had poor social participation: 16 cases (84.2%) were males and 3 females (15.8%) 15 cases (40.5%) had poor school performances: 12 cases (80%) were males and 3 females (20%).

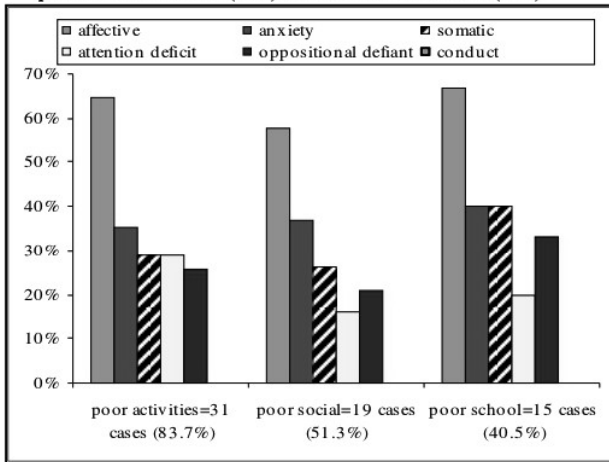


Figure (4) Bar chart illustrating behavior problems percentage in relation to activities, social and school performance.

Table (4) Behavioral problems in relation to activities participation:

Behavioral Problems	Activities participation (N= 37)		R	P Value	
	Poor Activities (31 Cases)	Normal Activities (6 Cases)			
	No (%)	No (%)			
Affective Problems	Yes	20 (64.5)	0 (0.0)	0.477	0.004
	No	11 (35.5)	6 (100.0)		
Anxiety Problems	Yes	11 (35.5)	0 (0.0)	0.286	0.082
	No	20 (64.5)	6 (100.0)		
Somatic Problems	Yes	9 (29.0)	0 (0.0)	0.249	0.129
	No	22 (71.0)	6 (100.0)		
Attention Deficit	Yes	3 (9.7)	1 (16.7)	-0.083	0.614
	No	28 (90.3)	5 (83.3)		
Oppositional Defiant Problems	Yes	8 (25.8)	0 (0.0)	0.231	0.160
	No	23 (74.2)	6 (100.0)		
Conduct Problems	Yes	2 (6.5)	0 (0.0)	0.105	0.522
	No	29 (93.5)	6 (100.0)		

The affective problems were significantly correlated with the poor activities participation and correlation coefficient was moderately positive. The other behavioral problems showed insignificant correlations with the poor activities.

Table (5) Behavioral problems in relation to social participation:

Behavioral Problems	Social participation (N= 37)		R	P Value	
	Poor Activities (31 Cases)	Normal Activities (6 Cases)			
	No (%)	No (%)			
Affective Problems	Yes	11 (57.9)	0 (0.0)	0.633	0.000
	No	8 (42.1)	18 (100.0)		
Anxiety Problems	Yes	7 (36.8)	0 (0.0)	0.470	0.004
	No	12 (63.2)	18 (100.0)		
Somatic Problems	Yes	5 (26.3)	0 (0.0)	0.385	0.019
	No	14 (73.7)	18 (100.0)		
Attention Deficit	Yes	3 (15.8)	1 (5.6)	0.165	0.316
	No	16 (84.2)	17 (94.4)		
Oppositional Defiant Problems	Yes	4 (21.1)	0 (0.0)	0.339	0.039
	No	15 (78.9)	18 (100.0)		
Conduct Problems	Yes	2 (10.5)	0 (0.0)	0.233	0.157
	No	17 (89.5)	18 (100.0)		

blems, somatic problems and oppositional defiant problems were significantly correlated with the poor social participation.

Table (6) Behavioral problems in relation to school performance

Behavioral Problems	School performance (N= 37)		R	P Value	
	Poor Activities (15 Cases)	Normal Activities (22 Cases)			
	No (%)	No (%)			
Affective Problems	Yes	10 (66.7)	0 (0.0)	0.737	0.000
	No	5 (33.3)	22 (100.0)		
Anxiety Problems	Yes	6 (40.0)	0 (0.0)	0.533	0.001
	No	9 (60.0)	22 (100.0)		
Somatic Problems	Yes	6 (40.0)	0 (0.0)	0.533	0.001
	No	9 (60.0)	22 (100.0)		
Attention Deficit	Yes	3 (20.0)	1 (4.5)	0.244	0.137
	No	12 (80.0)	21 (95.5)		
Oppositional Defiant Problems	Yes	5 (33.3)	0 (0.0)	0.479	0.004
	No	10 (66.7)	22 (100.0)		
Conduct Problems	Yes	2 (13.3)	0 (0.0)	0.289	0.078
	No	13 (86.7)	22 (100.0)		

The affective problems, anxiety problems, somatic and oppositional defiant problems were significantly correlated with the poor school performance of SEMDs cases.

The comparison of the results of different behavioral problems in relation to gender of SEMDs patients were illustrated in figure (5) and table (7).

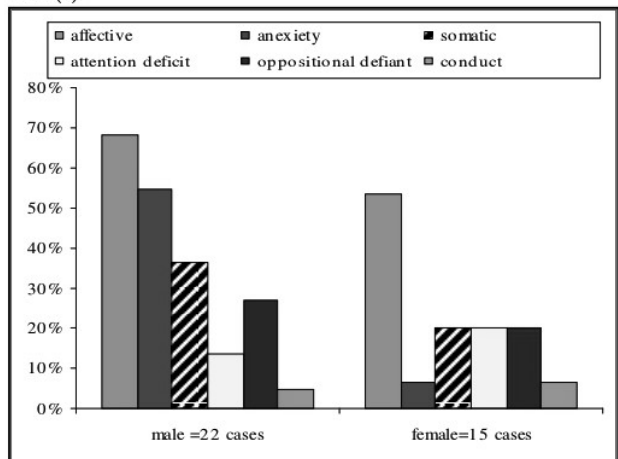


Figure (5) Bar chart illustrating behavioral problems percentage in relation to the sex of the studied SEMDs patients.

Table (7) Behavioral problems in relation to gender

Behavioral Problems		Gender (N= 37)		R	P Value
		Male= 22 Cases	Female= 15 Cases		
		No (%)	No (%)		
Affective Problems	Yes	15 (68.2)	8 (53.3)	0.15	0.361
	No	7 (31.8)	7 (46.7)		
Anxiety Problems	Yes	12 (54.5)	1 (6.7)	0.492	0.003
	No	10 (45.5)	14 (93.3)		
Somatic Problems	Yes	8 (36.4)	2 (13.3)	0.255	0.121
	No	14 (63.6)	13 (86.7)		
Attention Deficit	Yes	3 (13.6)	4 (26.7)	- 0.163	0.320
	No	19 (86.4)	11 (73.3)		
Oppositional Defiant Problems	Yes	6 (27.3)	3 (20.0)	0.083	0.613
	No	16 (72.7)	12 (80.0)		
Conduct Problems	Yes	1 (4.5)	1 (6.7)	- 0.046	0.779
	No	21 (95.5)	14 (93.3)		

The correlation between gender and behavior problems showed that all scales were insignificantly related to gender except anxiety problems which were significantly common in male patients compared to female patients and correlation coefficient was moderately positive.

Discussion:

SEMDs are a heterogeneous group of skeletal dysplasias. Patients affected with SEMDs usually have variable degrees of short stature and skeletal deformities of the spine and long bones. Management of these dysplasias needs multidisciplinary team. The evaluation of the psychological burden which may be associated with SEMDs is important for further management and psychiatrist referral.

From review of literature, little studies were discussed the psychological burden of short stature associated with skeletal deformities on patients with skeletal dysplasias. Few studies were discussed that effect on patients of achondroplasia and osteogenesis imperfecta. Up to our knowledge, no studies were discussed the psychological effect of short stature and skeletal deformities which associated with patients of SEMD till now. This study is the first study to assess the behavior problems which may accompany the skeletal deformities associated with SEMDs by using the child behavior checklist.

Our study showed that most of our patients (62.1%) had more liability or clinical range to affective problems (dysthymia and depression) than other studied behavior problems. Anxiety problems were the next (35.1%), followed by somatic, oppositional defiant and attention deficit, respectively. Two patients showed a liability to conduct problems.

Our study showed that all the behavioral scales were significantly higher than general population and mean T scores of SEMDs patient of all scales was 61.1±7.46 (p value= 0.002). The mean T score of internalizing behavioral scales was higher than externalizing scales (65.63±7.58: 56.58±7.34). The mean T score of internalizing scales is considered borderline and the mean T score of affective problems of SEMDs patients is considered clinically concerning compared to the general population (mean= 71.43± 8.54). Our results showed that SEMDs patients had more liability to internalizing behavioral problems in comparison to the general population specially the affective problems as mean T score of our patients

were within the clinical range although all subscales were high.

An Australian descriptive study of 53 children with achondroplasia (the most common skeletal dysplasia) and an age between 5 and 17 years was done by Wigg et.al, (2016). While their results indicated generally low levels of difficulties on the CBCL and the presence of some thought problems, and anxiety problems. Our study showed that cases with SEMDs were more vulnerable to behavioral problems in comparison to the Australian study of Achondroplasia patients, although both are a subgroup of skeletal dysplasia but it seems that Achondroplasia patients were more adapted. Maybe the difference of the disease onset, manifestations and demographic factors as the different societies ought to be considered to explain our results.

Random- effects meta- analysis was computed to integrate the results of 569 studies that used the CBCL to examine the risk of emotional and behavioral problems among children with a chronic different physical illness. It found that children with a chronic physical illness have higher levels of internalizing, externalizing and total behavior problems than healthy peers. Effects also varied by country and, in part, by age and gender. It found that the strongest effect sizes for internalizing problems were found with those illnesses that are associated with a higher number of stressors (such as activity restrictions, frightening symptoms, and lack of control over symptoms). The strongest levels of externalizing problems were found with illnesses that affect brain function (epilepsy, headache, intellectual disability and spina bifida) (Pinquart& Shen, 2011).

Most of our SEMDs cases had a higher numbers of stressor as the 1st group of the computed meta- analysis of Pinquart& Shen, (2011) and most of our patient similarly had significantly associated internalizing problems.

The comparison of the results of different behavioral problems of the studied thirty seven patients with SEMDs of different degrees of skeletal severity showed no significant difference between mild, moderate and severe cases in the prevalence of behavioral problems. However, a study with larger patient sample may be needed to provide more definitive results.

The correlation of IQ grade with behavioral problems showed that SEMD patients who had IQ> 70 had a higher percentage of externalizing problems than internalizing problems although p value were insignificant. Very low IQ cases showed more significant liability or clinical range for externalizing problems. However, a study with larger patient sample may be needed to provide more definitive results. These results were matched with the results of the computed meta- analysis of Pinquart& Shen, (2011) as they found that chronic illnesses that affect brain function had more externalizing problems. However, a study with larger patient sample may be needed to provide more definitive results.

Activities, social and school performance were assessed by the use of the CBCL Competence Scales to study the underlying factors of our behavioral problems results. Results showed that the activities participation was more affected in our patients followed by the social participation then school participation, respectively. Males' percentage

was more than females' percentage.

The correlation between the studied behavioral problems and the results of Competence Scale showed that affective problems were significantly associated with poor activities (p value= 0.004) and R was moderately positive ($R= 0.477$). These results indicate that poor activities participation, poor social participation and the poor school performance may be the underlying causes of behaviors changes in patients with SEMDs. The higher percentage of poor activities in our studied patients (83.7%) might be associated with their limited painful mobility but also may be due to the under practice of sports which is a common trend in the Egyptian population.

The affective problems, anxiety problems, somatic problems and Oppositional defiant problems were significantly correlated with the poor social participation (p value= 0.001, 0.004, 0.019 and 0.039 respectively) and correlation coefficient was moderately positive ($R= 0.633, 0.470, 0.385$ and 0.339 respectively). The other behavioral problems showed insignificant p values.

The affective problems, anxiety problems, somatic and Oppositional defiant problems were significantly correlated with the poor school performance of SEMDs cases (p value= 0.001, 0.001, 0.001 and 0.004 respectively) and correlation coefficient was moderately positive (correlations coefficient= 0.737, 0.533, 0.533 and 0.479 respectively). Attention deficit and conduct problems showed insignificant p values.

From our results poor activities, poor social and poor school performances could be considered an underling risk factors for our internalizing problems results which also matched with the results of the computed meta- analysis of Pinquart& Shen, (2011).

However, our results were not matching with the review of psychosocial study of children living with osteogenesis imperfecta OI (a group of skeletal dysplasia with an associated repeated fractures and deferent degrees of skeletal deformities and short stature) which found that cases with OI had good psychosocial adaptation. They had the ability of learning new skills, making social relationships and performing well in their schools although their limited mobility, fear of fractures and frequent school absence (Tsimicalis et.al., 2016). Both SEMDs and OI patients experience more or less similar manifestations although they adapt difference. That highlights the needs of more study to understand the underlying causes of that difference.

The correlation between gender and behavior problems showed that the affective problems, somatic and the oppositional defiant were insignificantly common in male patients. Anxiety problems were significantly common in male patients compared to female patients (p value= 0.003) and correlation coefficient was moderately positive (correlation coefficient= 0.492). our results showed that both gender were suffering as the diffrence between both was insignificant and males were significantly more suffering only with anxiety. In a larger study, Sandberg et. al. (1994) identified some difficulties of behavioral adaptation, mainly in males with short stature which little matches with our results.

Review of literature was inconclusive regarding whether skeletal dysplasias in general and the associated skeletal deformities and short statures were reasons of different behavior problems or not. Adelson (2005) reviewed previous studies of the medical and psychological aspects of profound short stature and associated skeletal dysplasia and concluded that those studies were done on different countries of western societies and found that persons with skeletal dysplasias were similar to average statured individuals with a little bit liability to anxiety and depression with some individuals doing poorly for long periods and many others experiencing significant ups and downs in response to situational challenges.

Short stature constitutes a psychosocial burden is a widespread belief. In a survey which was reported by Voss and Sandberg (2004), 56% of physicians supported that height impairs the emotional well- being in children when below the 3rd centile. Thirty two percent of these same physicians also believed that quality of life in short stature could be improved by an increase in height.

Association of short stature due to skeletal dysplasia with psychological outcomes was studied by Wheeler et.al. (2003) and they found no evidence for increased rates of depression or anxiety in children with skeletal dysplasia. Although, they recommended further studies to accurately evaluate psychological problems such as depression and anxiety to validate their results.

An older study by Skuse et.al. (1994) reported no significant differences in peer acceptance, self- perception or social competence between clinically referred short children and their classmates.

Although, not all children with short stature are referred for specialist evaluation. Those seeking help are likely to have greater problems of psychosocial adaptation than those who do not. Parental perceptions are also potentially biased and any behavioral or emotional problems in the child may well be interpreted as stature related, even where this is not the case.

In our study, we found that there is a significant association between the skeletal dysplasias and behavior problems specially the affective behavior problems (62.1%) and that the internalizing problems were significantly associated with SEMDs.

The severity of short stature and skeletal deformities did not significantly affect the behavioral problems prevalence.

Cases with $IQ < 70$ had more externalizing problems than cases with better IQ grades.

Patients of SEMDs had high percentage of poor activities participating, social participation and school performance which could be considered an underling factors to their poor psychological adaptation. However, more social and cultural aspects should be studied to evaluate the cause of this significant difference.

Both gender were suffering except for anxiety problems males were poorly adapted and more suffering

Our study supports that short stature constitutes a psychosocial burden

and impairs the emotional well-being of SEMD patients. Stigmatization, victimization, poor self-perception, parents' overprotection, poor school achievement due to school unattendance, all can lead to immature personality and may be the causes of behavior problems.

Referral of our studied cases to a psychologist was done for further and suitable management. Group psychotherapy construction of cases with SEMDs also was recommended. Follow up and reassessment are very important.

Conclusion:

SEMDs are associated with behavior changes. The affective problems were the most common changes with SEMDs. The severity of SEMDs complications was not correlated with more affective changes. Males were more affected than females with anxiety. Poor activities, social and school performance may be the underlying causes for the behavior changes associated with SEMDs patients. The affected SEMDs cases need early diagnosis for further referral and early management.

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