# A Study of developmental vertebral disorders among Egyptian children

Thesis Submitted for Partial Fulfillment of the Master Degree in Pediatrics 20175 C. V.



By

Lerine Bahy El - Din El-Shazly

M.B.B.Ch.

**Supervisors** 

Prof. Dr. Rabah M. Shawky

Professor of Pediatrics & Genetics Faculty of Medicine Ain Shams University

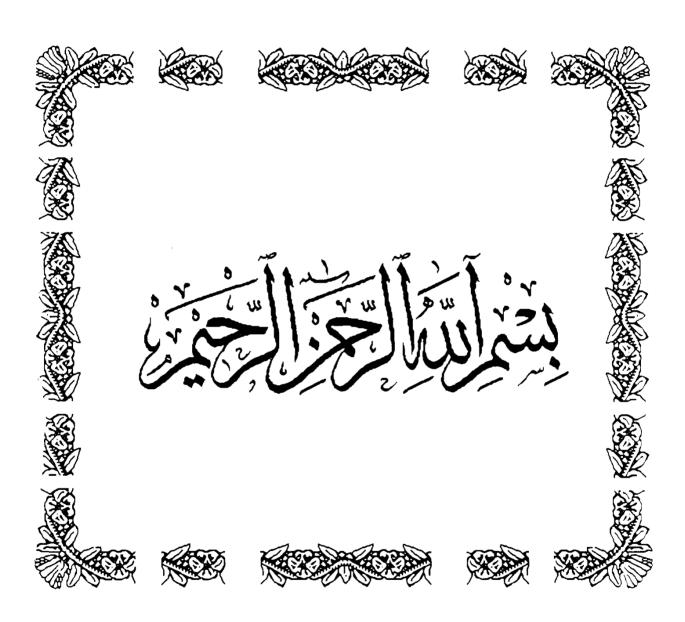
Dr. Eman Ahmed Zaky

Lecturer of Pediatrics Faculty of Medicine Ain Shams University

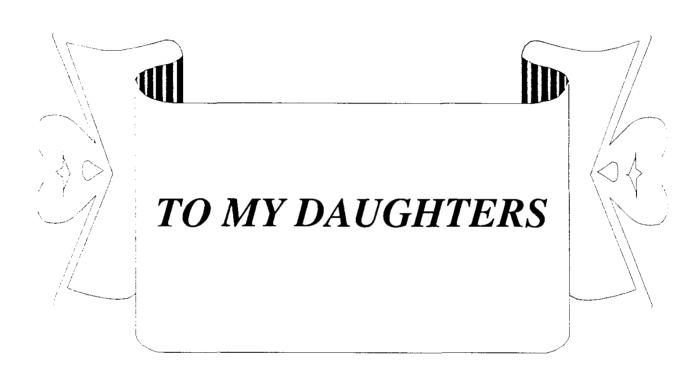


Faculty of Medicine Ain Shams University

1995







#### ACKNOWLEDGEMENT

First and foremost thanks are to ALLAH "The most beneficent, the merciful".

Second, I would like to express my deepest thanks and gratitude to Prof. Dr. RABAH M. SHAWKY, Professor of Pediatrics and Genetics, Faculty of Medicine, Ain Shams University, for her instructive advice and continuous support. Moreover, I sincerely appreciate her valuable time, effert and understanding. No words can express my feelings and respect for her.

Appreciative thanks to Dr. EMAN AHMED ZAKY, Lecturer of Pediatrics, Faculty of Medicine, Ain Shams University, for her valuable advices, scientific guidance, her fruitful discussion and for the continuous assistance during this work.

I would like also to express my appreciation to the help and cooperation, I have recieved from Dr. AMER KHAIRY ABD-EL-AZIZ, Lecturer of Orthopaedics, Faculty of Medicine, Ain Shams University.

I wish to express my sincere thanks to the patients and their parents included in this work for their greatful cooperation with me.

Finally, I appreciate the effort done by many members of Radiology Unit at Pediatric Hospital, Faculty of Medicine, Ain Shams University in production of this work.

## **CONTENTS**

CHAPTER	PAGE
INTRODUCTION & AIM OF THE WORK	1
REVIEW OF LITERATURE	3
I-Embryology and anatomy of the vertebral column	3
II- Classification of spinal disorders	21
1- Idiopathic Conditions	27
2- Neuromuscular Disorders	30
3- Congenital Disorders	39
4- Neurofibromatosis	61
5- Mesenchymal Diseases	62
6- Ostechondrodystrophies	68
7- Scheuermann's Disease	74
8- Metabolic Disorders	76
9- Conditions related to Lumbosacral area	81
10- Hysteria	85
11- Functional Disorders	86
III- Evaluation of patients with spinal deformities	88
1-History taking	88 89
2- Physical Examination	93
3- Radiologic Evaluation	
IV- Treatment of spinal deformities	107
SUBJECTS AND METHODS	121
	123
RESULTS	214
DISCUSSION	224
SUMMARY AND CONCLUSIONS	
RECOMMENDATIONS	226
	227
REFERENCES	

# List Of Figures

		Page
Fig (1)	Scheme to show the formation of the vertebral column at various	5
	stages of development	
Fig (2)	Structure of the Vertebral Column	8
Fig (3)	Structure of A Vertebra	9
Fig (4)	Structure of Atlas and Axis	13
Fig (5)	Structure of Cervical vertebrae	13
Fig (6)	Structure of Thoracic vertebra (side view)	14
Fig (7)	Demonstrating the small size of a vertebral foramen	15
Fig (8)	Sacrum and Coccyx, pelvic surface and base	16
Fig (9)	Structure of Sacrum and Coccyx	17
Fig (10)	Diagram showing hemivertebra at T3	18
Fig (11)	Structure of intervertebral disc	18
Fig (12)	Types of Congenital scoliosis	40
Fig (13)	Types of Congenital kyphosis	46
Fig (14)	Features of Marfan's syndrome	63
Fig (15)	Enzymes in the pathway of methionine metabolism	65
Fig (16)	Features of Ehlers-Danlos syndrome	67
Fig (17)	Features of Osteogenesis imperfecta	77
Fig (18)	Features of Osteogenesis imperfecta (continued)	<i>78</i>
Fig (19)	Treatment of Osteogenesis Imperfecta by Intramedullary	80
	Fixation and Orthoses	
Fig (20)	Terminology used in describing the scoliotic curve.	95
Fig (21)	The Lippman - Cobb method of measuring the degree of scoliotic	96
	curvature	
Fig (22)	The Risser -Ferguson method of measuring the degree of	97
	scoliotic curvature	
Fig (23)	The measurement of scoliosis using the scoliotic index	98
Fig (24)	Cobb's spinous-process method for determining vertebral rotation	100
Fig (25)	Moe's pedicle method for determining rotation	101
Fig (26)	Determination of skeletal maturity from the status of ossification	102
	of the vertebral ring apophysis	
Fig(27)	Determination of skeletal maturity from the status of ossification of the iliac apophysis	103

Fig (28)	Etiological classif: ion of studied patients with developmental vetrebral anomaius	124
Fig (29)	Statistical coversion between different studied groups as	126
Fi (30)	regards measure age of presentation  Sex distribution of of studied patients with developmental vetrebral anomalies	127
(31)	Statistical comparison between different studied groups as regards sex distribution	128
Fig (32)	Main presenting symptoms in studied patients with developmental vetrebral anomalies	130
Fig (33)	Incidence of consanguinity among studied patients	132
Fig (34)	Statistical comparison between studied groups concerning consangiunity	133
Fig (35)	Incidence of + ve family history among studied patients	134
Fig (36)	Statistical comparison regarding family history between studied groups	135
Fig (37)	Incidence of exposure to antenatal hazards among studied patients	137
Fig (38)	Statistical comparison between studied groups regarding antenatal history	138
Fig (39)	Mode of delivery of studied patients	139
Fig (40)	Statistical comparison between studied groups regarding mode of delivery	140
Fig (41)	Incidence of significant postnatal history among studied patients	142
Fig (42)	Statistical comparison between studied groups regarding significant post-natal history	143
Fig(43)	Statistical comparison between different studied groups as regards mean weight of patients	145
Fig (44)	Classification of patients of group I (O.C.D.) according to WT on percentiale	146
Fig(45)	Classification of patients of group II (C.S.A.) according to WT on percentiale	147
Fig (46)	Classification of patients of group III (Misc) according to WT on percentiale	148
Fig( 47)	Statistical comparison between different studied groups as regards mean height of patients	150

		Page
Fig( 48)	Classification of patients of group I (O.C.D.) according to HT on percentiale	151
Fig (49)	Classification of patients of group II (C.S.A) according to HT on percentiale	152
Fig (50)	Classification of patients of group III (Misc) according to HT on percentiale	153
Fig (51)	Statistical comparison between different studied groups as regards mean span of patients	155
Fig (52)	Statistical comparison between different studied groups as regards mean upper segment of patients	156
Fig (53)	Statistical comparison between different studied groups as regards mean lower segment of patients	157
Fig(54)	Statistical comparison between studied groups regarding skull anomalies	159
Fig (55)	Types of skull anomalies among studied patients	160
Fig (56)	Statistical comparison between studied groups regarding abnormal somatic features	162
Fig (57)	Statistical comparison between studied groups regarding eye anomalies	163
Fig (58)	Statistical comparison between studied groups regarding ear anomalies	165
Fig (59)	Statistical comparison between studied groups regarding anomalies of nose and oral cavity	166
Fig (60)	Statistical comparison between studied groups concerning abnormalities of upper limbs	168
Fig (61)	Types of upper 'imb anomalies among studied patients	169
Fig (62)	Statistical comparison between studied groups concerning abnormalities of lower limbs	171
Fig (63)	Types of lower limb anomalies among studied patients	172
Fig (64)	Statistical comparison between studied groups as regards back anomalies	174
Fig (65)	Types of back anomains mong studied patients	175
Fig (66)	Statistical comparison octween studied groups regarding anomalies of cardiovascular system	177
Fig (67)	Statistical comparison between studied groups regarding chest anomalies	178

		Page
Fig (68)	Statistical comparison between studied groups concerning	180
	+ve abdominal findings	
Fig (69)	Statistical comparison between studied groups concerning	181
	genitourinary anomalies	
Fig (70)	Statistical comparison between studied groups regarding	183
	the incidence of backward	
Fig (71)	A case with Hunter syndrome	184
Fig (72)	A case with Morquio syndrome	185
Fig (73)	A case of hurler syndrome	186
Fig (74)	A case of hurler syndrome	188
Fig (75)	Acase with Achondroplasia	190
Fig (76)	Another case with Achondroplasia	192
Fig (77)	A case of achondroplasia in a neonate	193
Fig (78)	A case with Arnold-chiari malformation	194
Fig (79)	Another case with Arnold-chiari malformation	195
Fig (80)	Three cases of Meningomyelocele	197
Fig (81)	Plain X-ray chest and heart of a case with hemivertebra and	198
	left sided scoliosis	
Fig (82)	A case of sacral agensis	199
Fig (83)	Aonther case of sacral agensis	201
Fig (84)	A case with spina bifida occulta with a tuft of hair on the	203
	back	
Fig (85)	A case ofspinal muscle atrophy	205
Fig (86)	A case of spinal muscle atrophy	207
Fig (87)	A case with Kleipel Feil syndrome	208
Fig (88)	A case with congenital hypothyrodism with mental	210
	retardation	
Fig (89)	A case of idiopathic left scoliosis	211
Fig (90)	Plain X-ray chest and heart of a case with idiopathic right	212
	sided scoliosis	
Fig (91)	A case with Ruhinstein Tavhi syndrome	213

## List of Tables

Table (1)	Classification of mucopolysaccharidoses	73
Table (2)	Etiological classification of studied patients with developmental	
	vertebral anomalies.	123
Table (3)	Statistical comparison between different studied groups as regards	
	mean age of presentation.	125
Table (4)	Statistical comparison between different studied groups as regards	
	sex distribution.	125
Table (5)	Main presenting symptoms in studied patients with developmental	
	vertebral anomalies.	129
Table (6)	Statistical comparison between different studied groups	
	concerning consangiunity.	131
Table (7)	Statistical comparison regarding family history between studied	
	groups.	131
Table (8)	Statistical comparison between studied groups regarding	
	ante-natal history.	136
Table (9)	Statistical comparison between studied groups regarding natal	
	history.	136
Table (10)	Statistical comparison between studied groups regarding	
	post-natal history.	141
Table (11)	Statistical comparison between studied groups regarding mean	
	weight of patients.	144
Table (12)	Statistical comparison between studied groups regarding weight	
	on percentiales.	144
Table (13)	Statistical comparison between studied groups regarding mean	
	height of patients.	149
Table (14)	Statistical comparison between studied groups regarding height on	
	percentiales.	149
Table (15)	Statistical comparison between studied groups regarding mean	
	span of patients.	154

Table (16)	Statistical comparison between studied groups regarding mean	
	upper segment of the body of patients.	154
Table (17)	Statistical comparison between studied groups regarding mean	
	lower segment of the body of patients.	154
Table (18)	Statistical comparison between studied groups regarding skull	
	anomalies.	158
Table (19)	Statistical comparison between studied groups regarding somatic	
	features.	162
Table (20)	Statistical comparison between studied groups regarding eye	
	anomalies.	162
Table (21)	Statistical comparison between studied groups regarding ear	
	anomalies.	16
Table (22)	Statistical comparison between studied groups regarding	
	anomalies of the nose and oral cavity.	164
Table (23)	Statistical comparison between studied groups regarding	
	abnormalities of upper limbs.	16
Table (24)	Statistical comparison between studied groups regarding	
	abnormalities of lower limbs.	17
Table (25)	Statistical comparison between studied groups regarding back	
	anomalies.	17.
Table (26)	Statistical comparison between studied groups regarding	
	anomalies of cardiovascular system.	17
Table (27)	Statistical comparison between studied groups regarding	
	anomalies of chest.	17
Table (28)	Statistical comparison between studied groups regarding positive	
	abdominal findings.	17
Table (29)	Statistical comparison between studied groups concerning	
	genito-urinary anomalies.	17
Table (30)	Statistical comparison between studied groups regarding mental	
	backward	18

### List of Abbreviations

CS Caesarean section

CSA Congenital spinal anomalies

OCD Osteochondrodystrophies

Mis. Miscellaneous

NSVD Normal spontaneous vaginal delivery

# INTRODUCTION & AIM OF THE WORK

#### INTRODUCTION

Congenital and developmental spinal deformities are common and potentially serious non traumatic musculo skeletal problems in children and adolescents. Deformities may involve the spine curvature, appearing as a frontal plane curvature—scoliosis and as sagittal plane curves of lordosis and kyphosis. Deformities may be static, whereas some progress with time and require therapy (Staheli, 1992).

Developmental disorders of the pediatric spine are a group of congenital malformations commonly referred to as spinal dysraphism.

The ge malformations are characterized by incomplete or absent fusion of midline mesenchymal, bony, and neural structures. They can be classified according to the presence or absence of an associated back mass (Byrd et al., 1991).

Beals and his associates (1993) reviewed 218 patients with congenital vertebral anomalies. They demonstrated that 61% of their patients had associated abnormalities affecting seven systems. The type of vertebral anomaly did not predict the location or type of associated abnormality.