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Uncommon and Rare Pediatric Syndromes Associated with Surgical Conditions in Iraqi Children

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1. Abstract

Background: Many uncommon and rare pediatric clinical syndromes including genetic and dysmorphic disorders are known to be associated with surgical conditions and require various types of surgical interventions.

Little is known about the pattern of surgical conditions associated with the uncommon and rare childhood syndromes in Iraq.

The aim of this paper to describe our experience with uncommon and rare childhood syndromes associated with surgical conditions observed in two tertiary referral centers in Baghdad.

Patients and methods: Twenty one unrelated patients (10 males and 11 females) with uncommon and rare syndromes associated with surgical conditions were observed. The patient's age ranged from 3 days to 18 years. The patients were observed during the period from 1994 to December 2019 in two tertiary referral teaching hospitals (University Hospital in Al-Kadhimiyia which was called later "Al-Kadhimiyia Teaching Hospital" and the Children Teaching Hospital of Baghdad Medical City).

Results: The twenty one patients had seventeen uncommon and rare syndromes associated with surgical conditions including ocular conditions (Cataracts and glaucoma), congenital heart disease, gastrointestinal surgical conditions (Imperforated anus, congenital primary cricopharyngeal achalasia, omphalocele and neonatal Hirschsprung disease), urologic surgical condition (Undescended testes) and orthopedic surgical conditions.

(Hip dislocation and multiple fractures), gynecologic ocular conditions (Hematocolpos which is caused by hemivaginal obstruction) and surgical conditions needing plastic surgeries.

Conclusion: This unique series of pediatric syndromes associated with surgical conditions included a new Iraqi variant of oculocerebrorenal syndrome, the cases number 35 and 36 of cutis laxa type II (Debre type) in the literature, the case number 52 of Ruprecht Majewski-Bosma syndrome in literature, the 58th case of Toriello-Carey syndrome in the literature, the case number 130 of Townes Brocks syndrome, the twenty eighth case of congenital Chevalier Jackson in the literature, the thirty fourth case of Goldberg Shprintzen syndrome and the fourth patient in the literature with the syndrome of microtia/anotia syndrome and congenital facial palsy without other abnormalities

2. Keywords: Pediatric syndromes; Surgical conditions; Iraq

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3. Introduction

Many pediatric clinical syndromes including genetic and dysmorphic disorders are known to be associated with surgical conditions and require various types of surgical interventions. We have previously reported our extensive pioneering experiences in the fields of clinical genetics, clinical dysmorphology and rare disorders in a plethora of publications [1-9].

Little is known about the pattern of surgical conditions associated with childhood syndromes. The aim of this paper to describe our experience with childhood syndromes associated with surgical conditions.

4. Patients and Methods

Twenty one unrelated patients (10 males and 11 females) with uncommon and rare syndromes associated with surgical conditions were observed. The patient's age ranged from 3 days to 18 years. The patients were observed during the period from 1994 to December 2019 in two tertiary referral teaching hospitals (University Hospital in Al-Kadhimiyia which was called later "Al-Kadhimiyia Teaching Hospital" and the Children Teaching Hospital of Baghdad Medical City).

All the patients were included in previous publications [10-26].

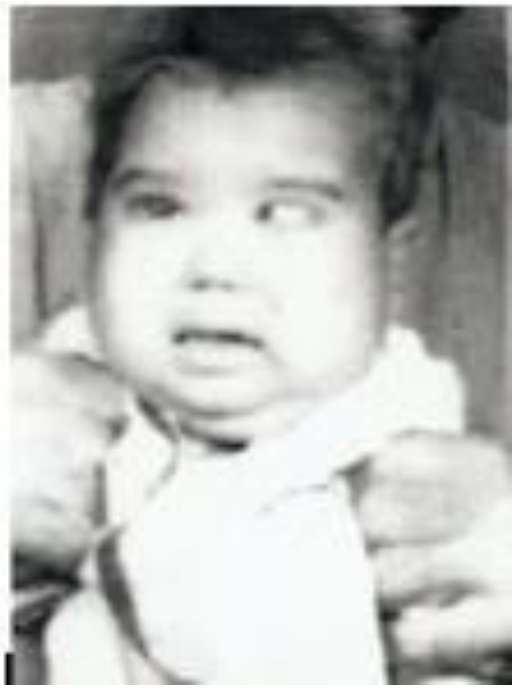


Figure 1A: A boy with an Iraqi variant of oculocerebrorenal syndrome. In addition to cataracts, he had developmental delay, hypotonia and hyporeflexia nystagmus and bilateral congenital chorioretinal hypoplasia.

Figure 1B: A girl with an Iraqi variant of oculocerebrorenal syndrome. She had bilateral congenital glaucoma and left cataract.

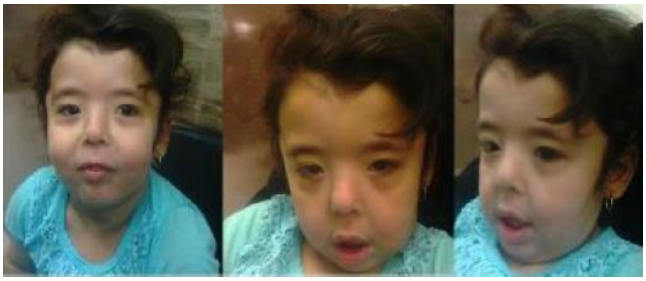
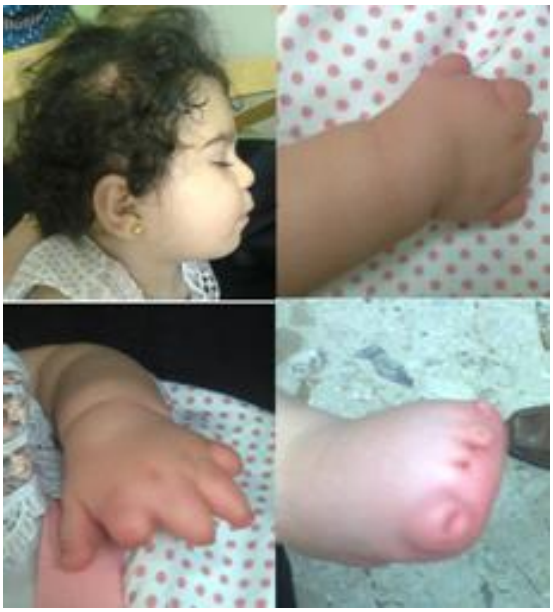


Figure 2: A girl with cutis laxa type II (Debre type) who had atrial septal defect.



Figures 3: The girl with Adams Oliver syndrome had terminal transverse limb and congenital scalp defect. She also had hairless regions in the scalp and low set ears.

5. Results

The twenty one patients had seventeen uncommon and rare syndromes (Table-1).

Five patients had congenital heart disease including a girl with cutis laxa type II (Debre type) (Figure-2) who had atrial septal defect [12], a girl with Adams Oliver syndrome (Figure-3) who had supra-valvular aortic stenosis [18], a girl with Ruprecht Majewski-Bosma syndrome who had atrial septal defect and bilateral aplasia of the nose needing plastic surgery (Figure-4) [24].

And two patient had multiple cardiac defects each had Noonan syndrome (Figure-5) [21] and Toriello-Carey Syndrome (Figure-6) [22].

Table 1: Twenty one unrelated patients (10 males and 11 females) had seventeen uncommon and rare syndromes associated with surgical conditions.

	Syndrome	**No**	**M**	**F**	**Surgical condition**
1	Oculocerebrorenal syndrome	4	2	2	Congenital glaucoma and cataracts
2	Cutis laxa type II (Debre type).	2		2	One girl had dislocated hip and One girl had atrial septal defect
3	Mostyn Embrey syndrome	1		1	Hematocolpos which is caused by hemivaginal obstruction
4	Chevalier Jackson syndrome	1		1	Congenital primary cricopharyngeal achalasia
5	Beckwith Wiedemann syndrome	1	1		Omphalocele
6	Pallister Hall syndrome	1	1		Imperforated anus
7	Berry-Treacher Collins syndrome	1	1		Microtia needing plastic surgery
8	Ekman-Lobstein syndrome	1	1		Multiple fractures
9	Adams Oliver syndrome	1		1	Supra-valvular aortic stenosis
10	Townes Brocks syndrome	1		1	Imperforated anus
11	The syndrome of congenital facial palsy and unilateral anotia.	1		1	Anotia needing plastic surgery
12	Noonan syndrome	1	1		Undescended testes, interatrial septum, small atrial septal defect and closing patent ductus arteriosus.

13	Toriello-Carey Syndrome	1		1	Congenital heart defects :small subaortic ventricular septal defect, small muscular ventricular septal defect, secondum atrial septal defect, mild pulmonary stenosis and mild coarctation of the aorta
14	Mowat Wilson syndrome	1	1		Congenital dislocation of the hip
15	Ruprecht Majewski-Bosma syndrome	1		1	Bilateral aplasia of the nose needing plastic surgery to build an external nose Atrial septal defect
16	Extended Michelin tire baby syndrome	1	1		Undescended testes
17	Goldberg Shprintzen syndrome	1	1		Neonatal intestinal obstruction attributed to Hirschsprung disease

Figure 4: The girl with Ruprecht Majewski-Bosma syndrome who had atrial septal defect and bilateral aplasia of the nose needing plastic surgery.

Figure 5: The infant with Noonan syndrome had undescended testes and cardiac defects. He also had facial dysmorphic features consisting of low set ears, hypertelorism and smooth philtrum.

Two patients had imperforated anus each had Pallister Hall syndrome (Figure-7) [5] and Townes Brocks syndrome (Figure-8) [10].

Figure 6: The girl with Toriello-Carey syndrome who had congenital heart defects. She experienced neonatal seizures and had facial dysmorphism including telecanthus, narrow and short palpebral fissures, low set ears and retrognathia.

Gastrointestinal surgical conditions were also observed in another three patients including a girl with Chevalier Jackson syndrome who had congenital primary cricopharyngeal achalasia, a boy with Beckwith Wiedemann syndrome who had Omphalocele (Figure-9) [15] and a boy with who Goldberg Shprintzen syndrome (Figure-10) who had neonatal intestinal obstruction attributed to Hirschsprung disease [26].

Hip dislocation was observed in two patients including a girl with Cutis laxa type II (Debre type) (Figure-11) [11] and a boy with Mowat Wilson

syndrome (Figure-12) [23].

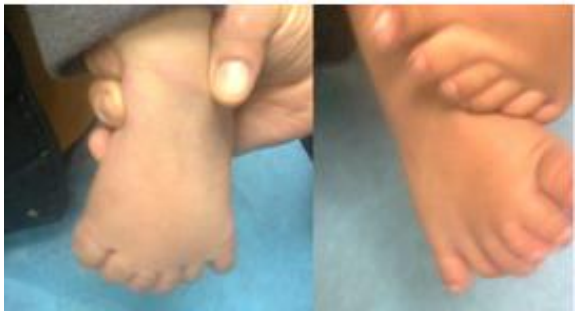


Figure 7: A boy with Pallister Hall syndrome who had imperforate anus at birth and polydactyly of hands and feet.

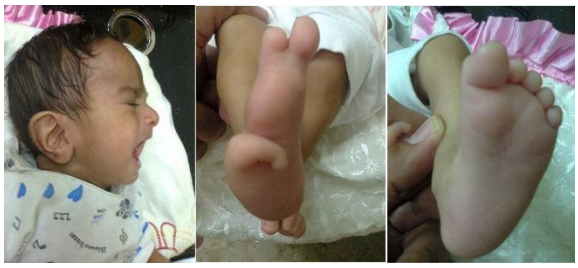


Figure 8: The girl with Townes Brock's syndrome and imperforated anus. She also had low set ears and deformity of the right foot with the presence of only three toes. There was no obvious abnormality of left foot, but the big toe was relatively large.

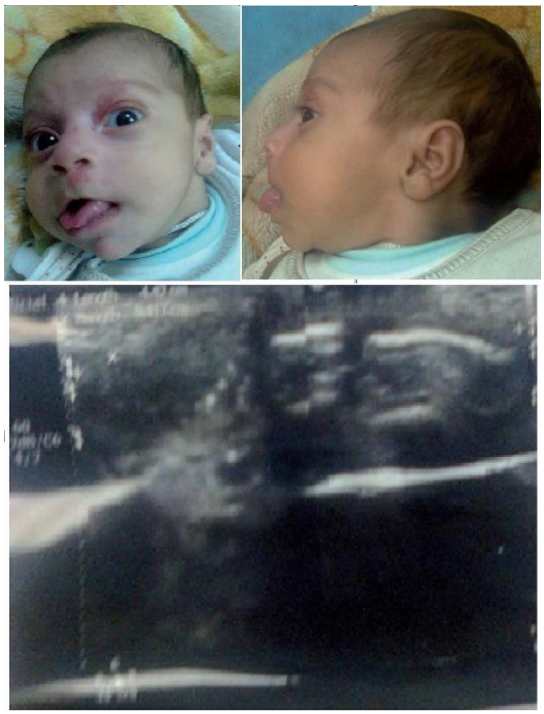


Figure 9: The boy Beckwith Wiedemann syndrome who had omphalocele. He also had recurrent hypoglycemia during infancy, macroglossia and nevus flammeus. Abdominal ultrasound before the operation showed evidence of abdominal wall defect at the center of the abdomen measuring 4.4 cm, reducible and containing bowel loops.

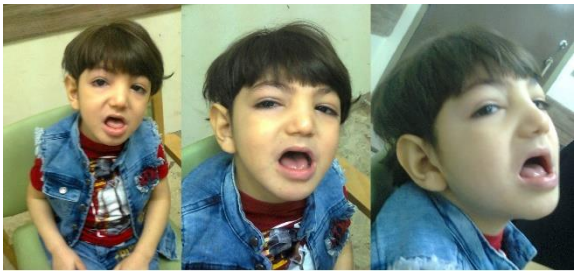


Figure 10: The boy with Goldberg Shprintzen syndrome who had neonatal intestinal obstruction attributed to Hirschsprung disease. He also had distinctive facial features characterized by hypertelorism, open mouth and laterally ear.

Another patient had orthopedic surgical condition "multiple fractures" was a boy who had Ekman-Lobstein syndrome (Figure-13) [12].

In addition, there were a girl with Mostyn Embrey syndrome had hematocolpos which is caused by hemivaginal obstruction, a boy with Berry-Treacher Collins syndrome had microtia (Figure-14) [16], a girl with syndrome of congenital facial palsy and unilateral anotia (Figure-15) [20].

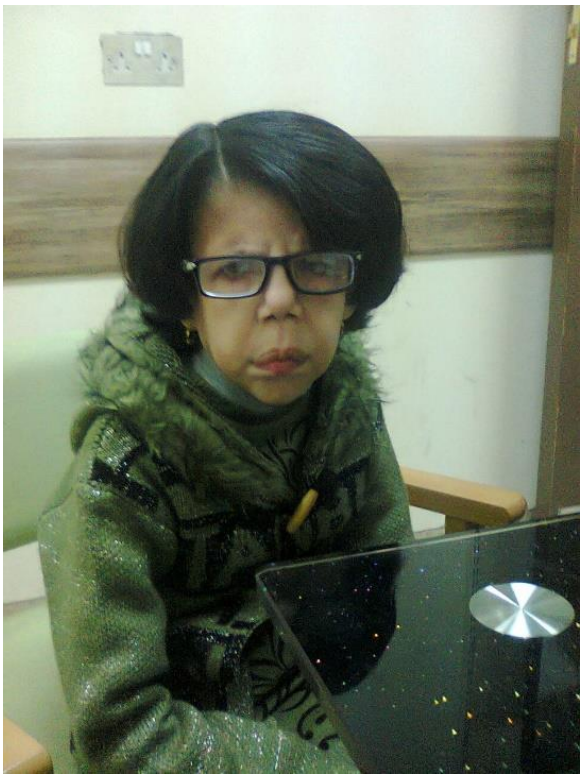


Figure 11: The girl with cutis laxa type II (Debre type) who had hip dislocation.

6. Discussion

We have previously described our extensive pioneering experiences with clinical genetic and dysmorphology in a plethora of publications [1-9].

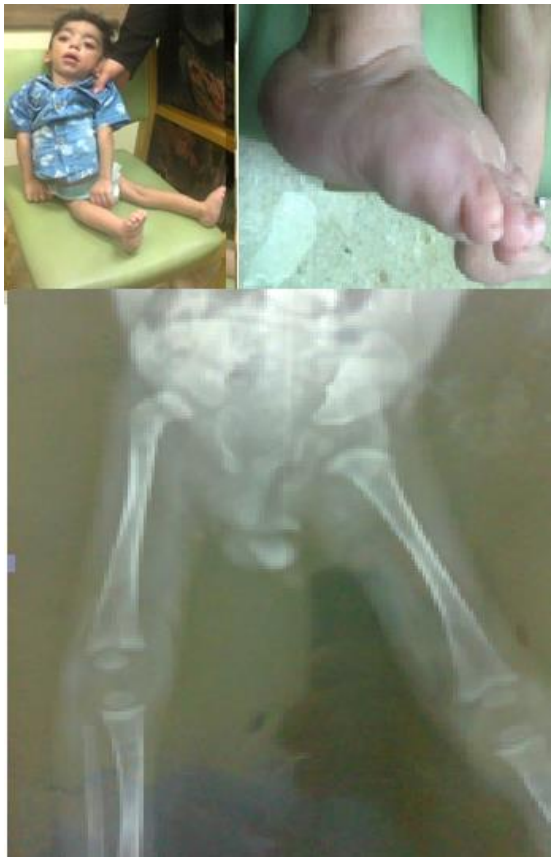


Figure 12: The boy with Mowat Wilson syndrome and hip dislocation. He had facial dysmorphism consisting of deep set large eyes, hypertelorism and open mouthed expression most of the time and low set ears. He also had bilateral rocker bottom feet and high arched palate. Radiographs showed advanced developmental dislocation of the right hip with superior lateral displacement and pseudo articulation of ileum.

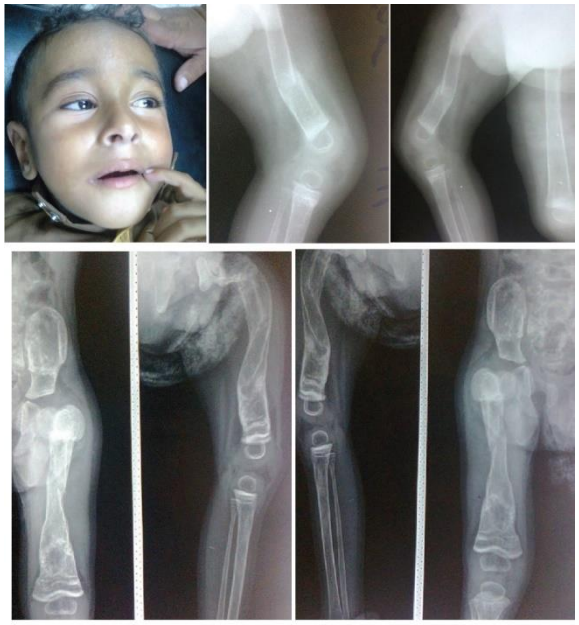


Figure 13: The boy with Ekman-Lobstein syndrome who had blue sclera and multiple recurrent fractures since early infancy. Radiographs showed osteoporotic changes, bowing of bones and multiple fractures.

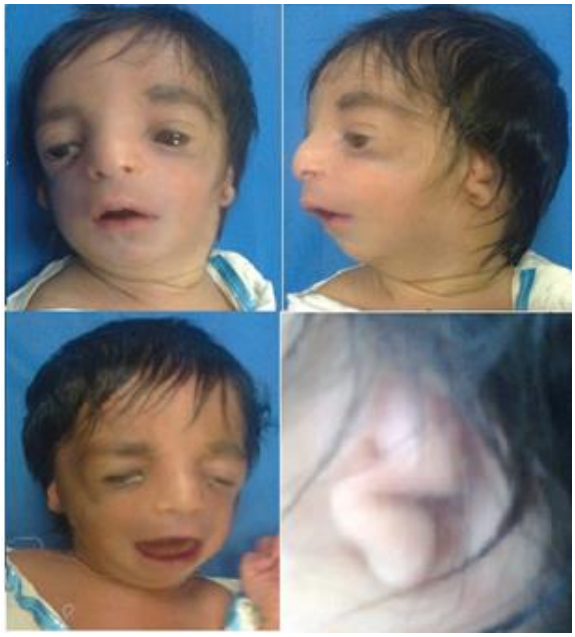


Figure 14: The boy with Berry-Treacher Collins syndrome. He had characteristic facial features including downward and laterally slanting palpebral fissures, paucity of lashes and lack of naso-frontal angle, bird like appearance, micrognathia, microtia (Deformed pinna), macrostomia and large tongue

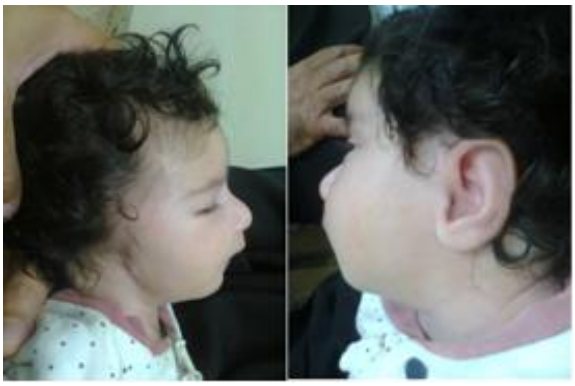


Figure 15: The girl with syndrome of congenital facial palsy and unilateral anotia. She had complete absence of the right ear.

This study included a unique series of rare and uncommon pediatric syndromes associated with surgical conditions including a new Iraqi variant of oculocerebrorenal syndrome, the cases number 35 and 36 of cutis laxa type II (Debre type) in the literature, the case number 52 of Ruprecht Majewski-Bosma syndrome in literature, the 58th case of Toriello-Carey syndrome in the literature, the case number 130 of Townes Brocks syndrome, the 28th case of congenital Chevalier Jackson in the literature, the thirty fourth case of Goldberg Shprintzen syndrome and the fourth patient in the literature with the syndrome of

microtia/anotia syndrome and congenital facial palsy without other abnormalities.

7. Conclusion

This paper represents a novel research describing rare and uncommon pediatric syndromes associated with surgical conditions in Iraqi children.

8. Acknowledgement

The author would like to express his gratitude for the parents of the patients who willingly accepted publishing the photos of their children and we also would to confirm the ownership of the copyrights of any previously published figure in this paper.

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