

GROWTH HORMONE RESISTANCE; IRAQI EXPERIENCE

Dr. Ali Hasan Al-Jumaili
FRCPCH, FACE, DCH, MBChB
Consultant paediatric endocrinology and diabetes
Member of the consultation panel of RCPCH for endocrinology and diabetes.
Representative of Iraq in the Arab society for paediatric endocrinology and diabetes(ASPED)

ePoster
code:P2-276



INTRODUCTION

Primary growth hormone resistance or growth hormone insensitivity syndrome (Laron syndrome) is an autosomal recessive disorder caused by deletions or mutations in the growth hormone receptor gene or by post receptor defect(1,2).

AIM

By presenting two Iraqi cases, with primary growth hormone resistance (Laron syndrome) we aim:
1.To highlight on the prevalence of this condition in Iraq. As a sample for more than twenty case detected.
2. Appeal to high health authorities and societies to help in providing and approving recombinant IGF-1 therapy for these patients .

CASE REPORT CON.

Physical examination: Both cases revealed cheerful and smart children with severe linear growth retardation. (Figures 1,2). The photos published according to consent of the families.

Clinical appearance of severe growth hormone deficiency :midfacial hypoplasia, frontal prominence, Saddle nose, flat nasal bridge High pitched voice, dental caries, and Poor dentition. (Figures 3,4,5).

Measurements: for the first case (female) ,weight 11 kg , height 79 cm both they were far below the 3rd centile (Z-score for height -10.6 SD) . For the second case(male), his weight 10 kg , height 72 cm (Z-score for height -7.6 SD).Figures :6,7,8,9.

Investigations: disclosed high plasma GH levels and low IGF-1 in both cases . The rest of the investigations were normal.



Figure 1:
first case
female
cheerful
and
smart

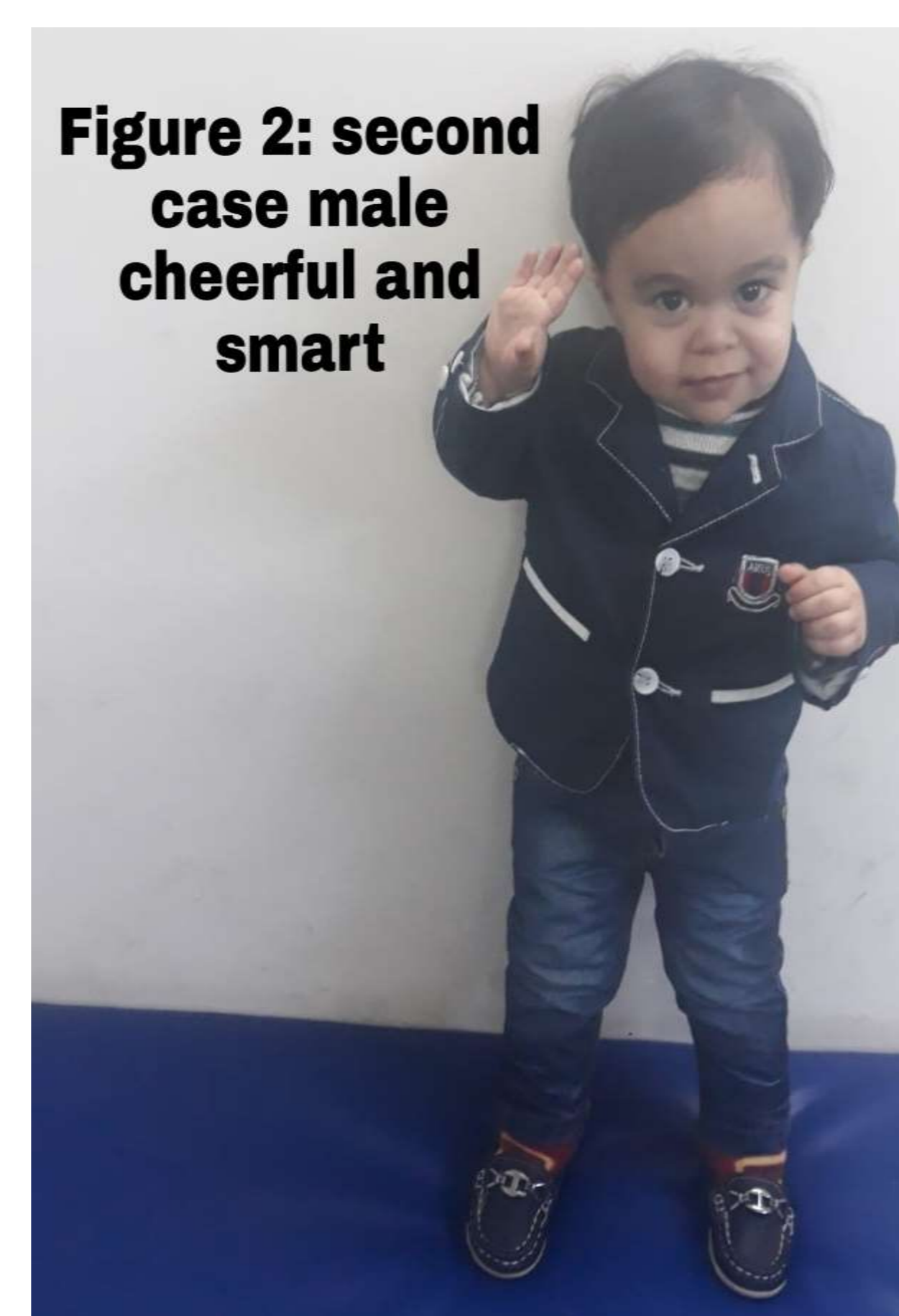


Figure 2: second
case male
cheerful and
smart

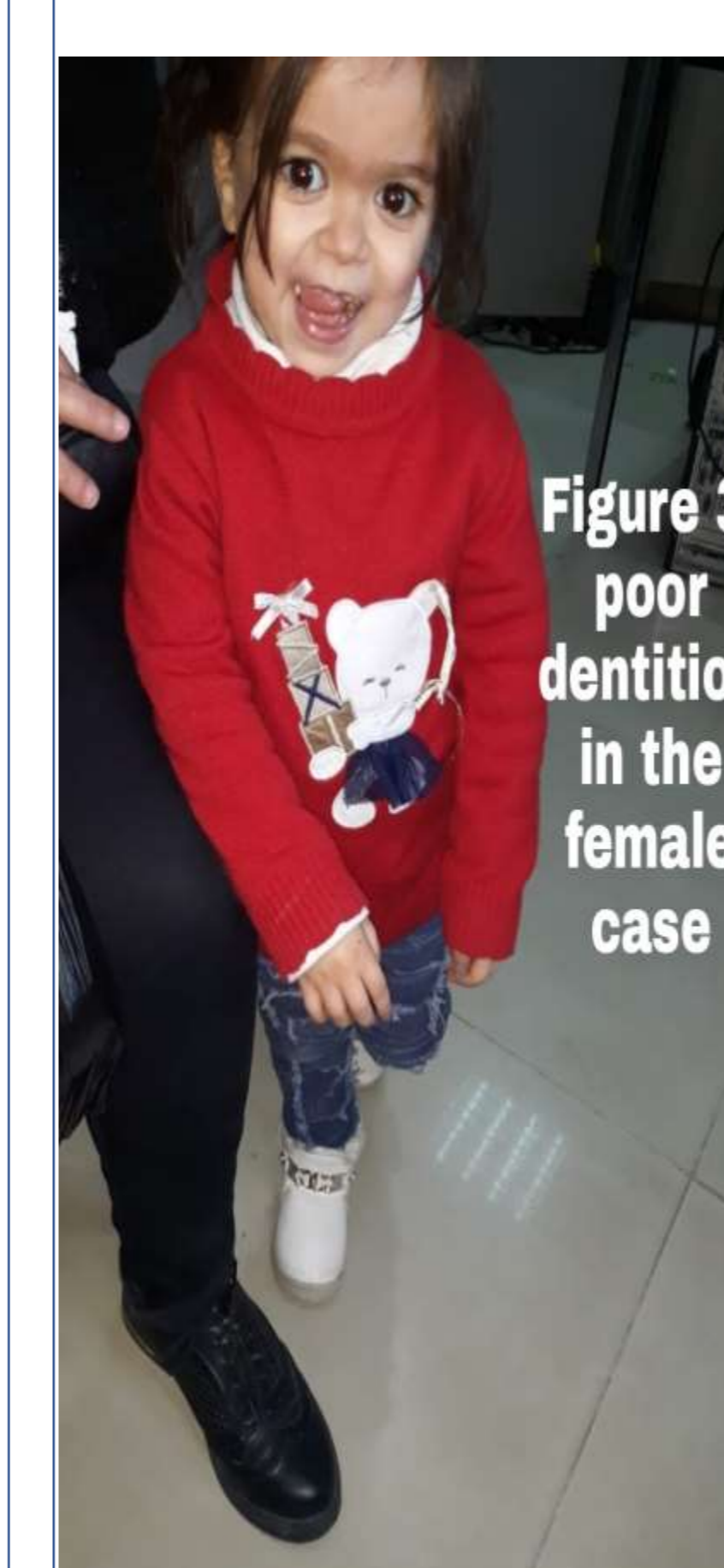


Figure 3:
poor
dentition
in the
female
case



Figure 4:
midfacial
hypoplasia,
frontal
prominence
, saddle
nose,flat
nasal
bridge in
female
case.



Figure 5:
midfacial
hypoplasia,
frontal
prominence
, saddle
nose,flat
nasal
bridge in
male case.

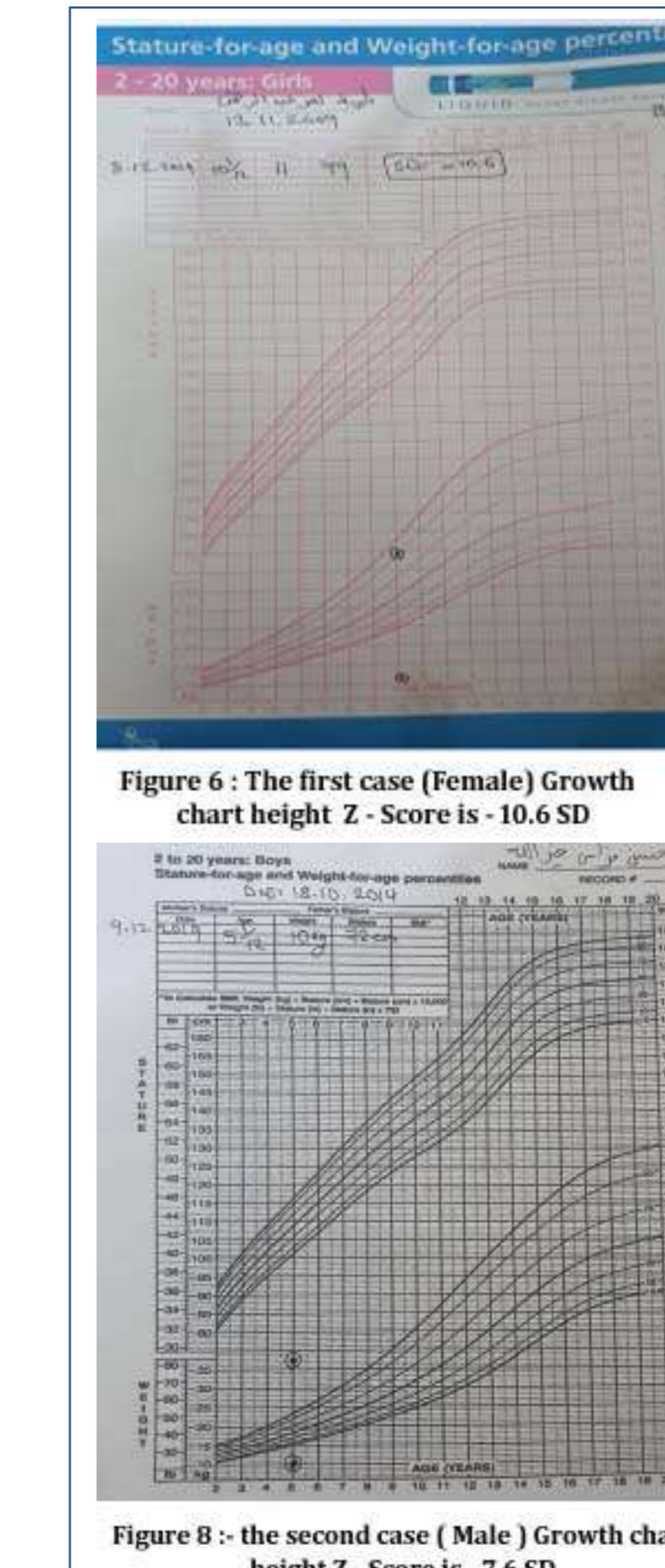


Figure 6 : The first case (Female) Growth chart height Z- Score is -10.6 SD



Figure 9 :-
Second case (Male)
with his peer of
the same age



Figure 7 : First case (Female)
with her peer of the same age 10 years

CASE REPORT

The first case : A ten years and two months old female child presented, with severe growth retardation neonatal hypoglycemic seizures. She was a product of caesarean section for consanguineous marriage. Her intrauterine period was uneventful; full term, birth weight was 3.750 kg .

The second case: A five years old male child who is the son for the cousin of the first case. Presented with postnatal growth failure. Consanguinity, positive for both families as for the patients. Birth weight 3.5 kg .

CONCLUSIONS

Conclusion: from clinical and laboratory findings are collectively consistent with primary growth hormone insensitivity (Laron Syndrome).(1,2). We conclude that:
1. Primary growth hormone resistance (insensitivity) or Laron syndrome is not a very rare condition as mentioned in the literatures(1,2,3). In Iraq, more than twenty case were detected. The same observation was also found in the Arab Gulf Countries who are members in ASPED.
2. The need for concerted efforts to provide and approved use of IGF-1 therapy for these patients is a paramount. Added to that the availability of genetic study .

REFERENCES

1. Clinical and Molecular Features of Laron Syndrome, A Genetic Disorder Protecting from Cancer
ANNA JANECKA, MARTA KOŁODZIEJ-RZEPA and BEATA BIESAGA
In Vivo July 2016, 30 (4) 375-381
2. AACE , Endocrine Practice REVIEW ARTICLES| VOLUME 21, ISSUE 12, P1395-1402, DECEMBER 01, 201
Lessons from 50 Years of Study of Laron Syndrome
Zvi Laron, MD, PhD(hc)
DOI:https://doi.org/10.4158/EP15939.RA
3. Human and Medical Genetics • Genet. Mol. Biol. 42 (4) • 2019 •
https://doi.org/10.1590/1678-4685-GMB-2018-0197 COPY
Growth Hormone insensitivity (Laron syndrome): Report of a new family and review of Brazilian patients
Thais R. VillelaBruna L. FreireNathalia T. P. BragaRodrigo R. ArantesMariana F. A. FunariJorge A L AlexanderIvanni N. Silva

ACKNOWLEDGEMENTS

No Acknowledgments

CONTACT INFORMATION

Email:alialjumaili27@yahoo.com
Mobile:00964 7901716132

