

# Incidence of ALS deficiency in patients with growth hormone deficiency at tertiary pediatric endocrinology center



K. RANKOVA<sup>1</sup>, V. IOTOVA<sup>1</sup>, Y. BOCHEVA<sup>2</sup>, S. SHEFKET<sup>2</sup>, N. YORDANOVA<sup>1</sup>, Y. BAZDARSKA<sup>1</sup>, K. TSOICHEV<sup>1</sup>, S. GALCHEVA<sup>1</sup>, V. BOYADZHIEV<sup>1</sup>

1. Department of Pediatrics, Medical University of Varna  
2. Department of General Medicine and Clinical Laboratory, Medical University of Varna

## INTRODUCTION

- Acid-labile subunit (ALS) is produced in the liver in response to growth hormone (GH)
- ALS forms a ternary complex with insulin-like growth factor I (IGF-I) and IGF binding protein-3 (IGFBP-3)
- ALS-deficient patients need specific therapeutic approach

## AIM

The aim of this study is to assess the prevalence of ALS deficiency in a cohort of patients with GH deficiency (GHD) followed up at a tertiary University pediatric endocrinology center.

## METHOD

- **71 children** (76% boys, age range 2-18 years)
- Diagnosed with GHD (auxology, 2 standard GH stimulation tests (max GH <10 ng/ml))
- Ongoing therapy with GH
- Collected blood serum samples were frozen at -80°C in 0.5 ml aliquots
- Standard ALS ELISA kit
- **11.6 ± 3.3 years** – the mean age of the children at the time of collection of samples

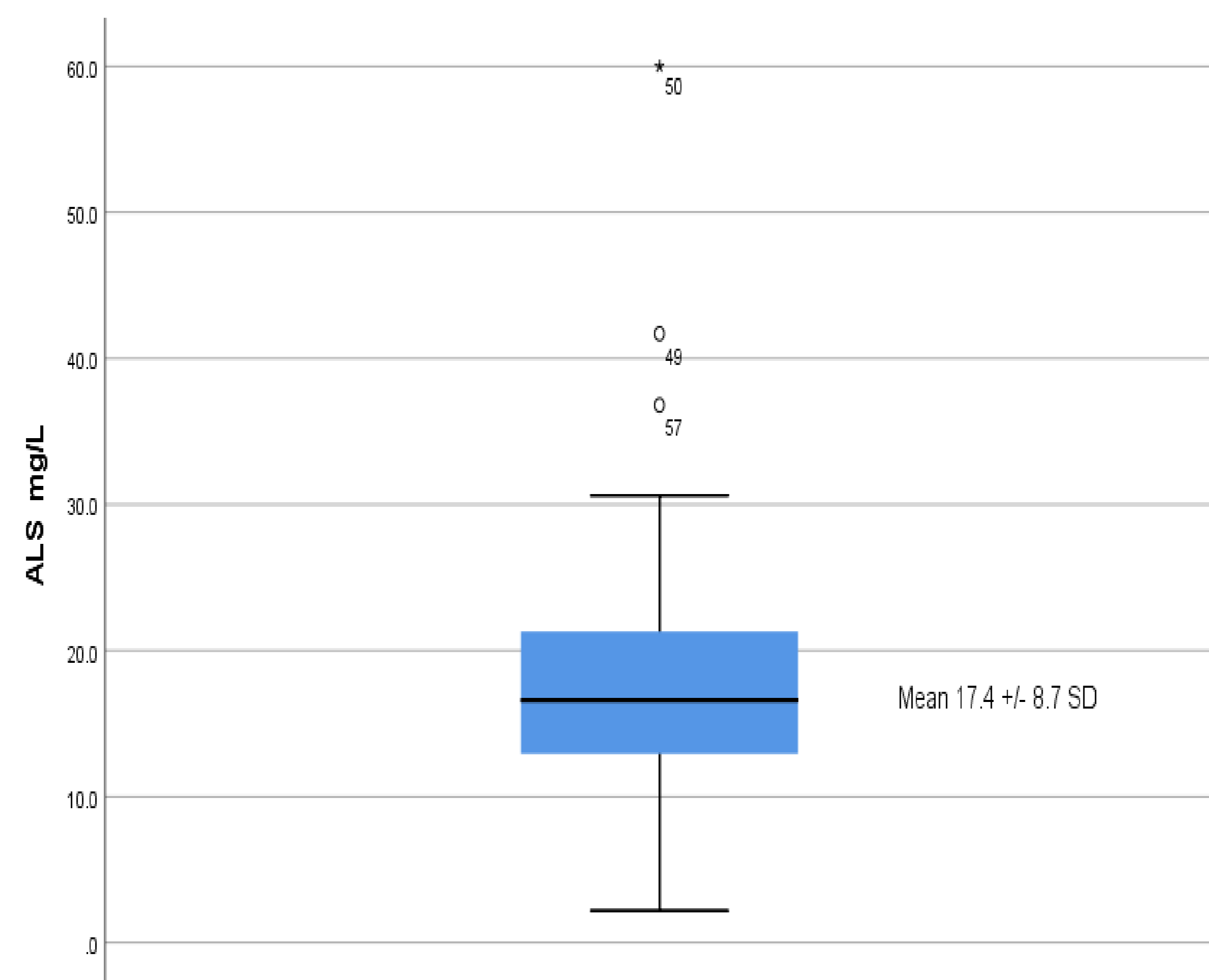
## RESULTS

### ALS deficiency screening:

- ✓ ALS levels range – 2.2 to 60 mg/L
- ✓ Mean - 17.4 ± 8.7 mg/L

### Published mean ALS levels from subjects without short stature:

- ✓ 24.2 ± 4.7 mg/L (2)
- ✓ 20.3 ± 3.1 mg/L (3)



	Mean ALS levels	Mean SDS height (before treatment)	Mean SDS IGF-1 (before treatment)
n	71	71	71
	17.4 ± 8.7 mg/L	-2.82 ± 1.21	-1.43 ± 1.0

➤ Low ALS levels correspond with low SDS<sub>height</sub> and low SDS<sub>IGF-1</sub> before therapy

3 patients with ALS levels < 4.0 mg/L

1 patient with ALS level 2.2 mg/L

close to those in patients with IGFBP3 gene mutations (<1.0 mg/L)

SDS height before therapy –3.1  
SDS IGF-1 before therapy –2.6  
Mean GH dose 1<sup>st</sup> y – 0.035 mg/kg/d  
SDS height after 1<sup>st</sup> y therapy – 2.1  
SDS IGF-1 after 1<sup>st</sup> y therapy – 0.67

## CONCLUSIONS

Our results show the prevalence of ALS deficiency in the current GH treated cohort and support the evidence that baseline investigation of ALS levels could be helpful in the differential diagnosis of growth disorders.

## REFERENCES

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Medical University Varna  
First Paediatric Clinic at UMHAT "St. Marina", Varna

## CONTACT INFORMATION

Kamelia Rankova  
Medical University Varna  
Bulgaria  
E-mail: kami.bachvarova@yahoo.com

