



## Correspondence

**Reply: Low bone mineral density is a common feature of Zellweger spectrum disorders**

**Keywords:**

Zellweger spectrum disorders  
 Bone health  
 Bone mineral density  
 Adrenal insufficiency  
 Glucocorticosteroid therapy

Dear Editor,

We read with interest the article of Rush et al. in which bone health in patients with a Zellweger spectrum disorder (ZSD) was investigated [1]. The authors determined the bone health of 13 patients with a ZSD through various tests, including biochemical analysis and a dual-energy X-ray absorptiometry (DXA) scan to assess bone mineral density (BMD). They show that most of the ZSD patients in this cohort have a significant low BMD for age. The authors hypothesize that this may be explained by activation of the peroxisome proliferator-activated receptor gamma (PPAR- $\gamma$ ) by very long chain fatty acids and phytanic acid, which consequently inhibits osteoblastogenesis.

Although this may be a contributing factor to the occurrence of low BMD in ZSD patients, we believe an important factor is left untouched by the authors. Since at least 30% of ZSD patients are diagnosed with adrenal insufficiency [2], it is essential to take into account how many patients in this cohort are chronically treated with glucocorticosteroids and in what dosage. Unfortunately, this is not mentioned in the article. Chronic use of glucocorticosteroids is widely known to have a negative impact on BMD as it stimulates the osteoclastogenesis and inhibits bone formation [3]. Even though the low BMD in ZSD patients is probably not solely caused by the chronic use of glucocorticoids, the extent to which this therapy could affect bone metabolism has to be taken into account.

Further research in which a distinction is made between ZSD patients with and without glucocorticoid therapy will therefore provide further understanding of the underlying pathology of low bone mineral density in ZSD patients.

**Abbreviations**

ZSD	Zellweger spectrum disorder
DXA	Dual-energy X-ray absorptiometry
BMD	Bone mineral density

**Competing interests**

The authors declare that they have no competing interests.

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**References**

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