

## INTRODUCTION

Diencephalic syndrome (DS), diencephalic cachexia or Russell syndrome, is a rare and rapidly fatal condition, usually occurring during the first year of life, as a result of a hypothalamic dysfunction due to hypothalamic/chiasmatic tumors.

Clinical features of DS are weight loss leading to cachexia despite a normal caloric intake and growth rate, hyperalertness, hyperkinesis, euphoria. Treatment is related to treatment of the hypothalamic lesion.

The role of cytokines, tumor-derived compounds, peptides and/or neuropeptides, neurotransmitters and hormones is still debated.

## OBJECTIVE AND HYPOTHESES

The aim of the authors is to evaluate if a longer endocrinological follow-up is necessary after treatment.

## METHODS

8 pediatric patients, 4 M and 4 F (median age at diagnosis of 6.5 months, range 4–60) followed at Meyer Children Hospital for DS as a result of an hypothalamic tumor.

Surgical treatment was based on tumor location and extent.

Patients received 10 monthly courses of cisplatin and etoposide and nutritional support.

Weight, length, head circumference and baseline endocrine function (IGF1, TSH, T4, cortisol, prolactin, ACTH and ADH) was evaluated before and after therapy.

## RESULTS

Despite different baseline endocrine values, at standard follow-up (2 years after therapy) we did not find any clinic endocrinological abnormality.

Prolonging follow-up, endocrine dysfunction developed in 2/8 patients. One had diabetes insipidus and precocious puberty and the other had a short stature due to GH deficit.

## CONCLUSIONS

As previously reported, our endocrinological data did not reveal any significant trend or correlation with the therapy immediately after treatment. But a longer follow-up revealed endocrinological diseases.

We conclude that a longer follow-up is necessary not only to better define long-term effectiveness of this low-dose cisplatin–etoposide regimen in the recovery of DS patients with hypothalamic tumors, but also to be able to recognise endocrine deficits.