



# A Case of Increased Desmopressin Requirement following Growth Hormone Administration in a Patient with a History of Histiocytosis

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## Patient Demographics

9 year 6 month old Caucasian female.

## Clinical Presentation/ Diagnosis

The patient has been managed in the pediatric endocrine clinic for several years following the diagnosis of diabetes insipidus (DI) and langerhan cell histiocytosis (LCH) of pituitary. She has been receiving oral desmopressin (brand name) on a stable regimen with few dosage changes required. Current dose of desmopressin 1.2 mg total daily dose.

There is a concern about her decreased growth velocity (4 cm/yr).

The patient has pituitary deficiencies in growth hormone (GHD) and antidiuretic hormone.

## Past History

- Initial presentation with polyuria, polydipsia, nocturia, weight loss and failure to thrive at age 5 years. Diabetes mellitus ruled out. Water deprivation test not conclusive for DI.

- Treated for gastroesophageal reflux without improvement in symptoms

- No growth for 1 ½ -2 years prior to initial presentation

- Langerhan cell histiocytosis of pituitary diagnosed 6 months later

- Skeletal survey negative

- Desmopressin initiated for treatment of DI

- Treated with chemotherapy for 18 months duration

- Developed recurrence of the tumor at age 7 years

- Re-treatment with chemotherapy for an additional year of therapy

- No medications except desmopressin following completion of chemotherapy

## Evaluation

- Brain MRI: 4.2x5.7x9 mm homogenous enhancing lesion in the infundibulum and hypothalamic region; langerhan cell histiocytosis of the pituitary confirmed by biopsy

- Laboratory studies done prior to initiation of growth hormone (GH) therapy

Test	Result
IGF-1	67 (88-474 ng/mL)
GH (stimulated Arginine/Insulin)	2.7 ng/mL
Cortisol (stimulated)	27 ug/dL
Sodium	140 (137 -145 mmol/L)
Osmolality	304 (275-295 mOsm/kg)

- Laboratory studies done following start of GH and increased desmopressin dose

Test	Result
Sodium	139 (137-145 mmol/L)
Fasting glucose	97 (74-106 mg/dL)
Osmolality	284 (275-295 mOsm/kg)
Urine osmolality	503 (100-1000 mOsm/kg)
Cortisol (fasting)	4.9 (2.9-19.4 ug/dL)
TSH	1.58 (0.7-6.4 ug/dL)
Free T4	1.04 (0.8-2.00 ng/dL)

## Interventions

- Desmopressin therapy initiated for treatment of DI

- GH therapy was initiated for treatment of GHD one year after completing the second course of chemotherapy

- Patient developed breakthrough from desmopressin with increased thirst and urination 2 weeks after beginning GH

Event	GH Dose	Desmopressin dose
Start GH	0.3 mg/kg/wk	1.2 mg/day
2 wks	0.3 mg/kg/wk	1.8 mg/day
D/C GH for 2 wks	0 mg/kg/wk	1.4 mg/day
Restart GH	0.15 mg/kg/wk	1.4 mg/day
1 month later	0.22 mg/kg/wk	1.6 mg/day

- No further changes in desmopressin requirement have been noted after 4 additional months of therapy, with increase in growth velocity to 8 cm/yr

## Discussion/ Recommendations

LCH (formerly known as histiocytosis X) is a rare disorder, of unknown etiology, characterized by histiocytic proliferation. It may be a reactive disease caused by abnormal immune regulation (3). LCH can involve multiple or single organs. DI is a common finding in this condition, frequently occurring as a seemingly idiopathic finding before other lesions are identified. Pituitary deficiencies of the anterior pituitary are less common, with GHD seen most frequently (1). Children with DI are at risk for panhypopituitarism with growth and development problems. Careful monitoring needs to be done to assess for growth hormone deficiency.

Treatment varies with the extent of the disease. Chemotherapeutic agents, including prednisone, vinblastin, and methotrexate have been used successfully (3).

## Conclusions

There have been no cases of this phenomenon reported in the literature.

The cause of the increased desmopressin requirement has not been explained. Fluid shifts with the initiation of growth hormone therapy may play a role in this need. While it is well known that dose-dependent and transient edema may occur with growth hormone therapy, this patient did not exhibit edema. Additional information is required to determine if there is a direct relationship between growth hormone therapy and the increased desmopressin requirements found in this case.

## References

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