

Recurrent severe hyponatremia in a young man with hydrocephalus and normal osmoregulatory function



Tudor RM¹, Hannon AM¹, Tormey WT², Sherlock M¹, Thompson CJ¹

¹Academic Department of Endocrinology and Diabetes Beaumont Hospital/RCSI Medical School Dublin

²Department of Chemical Pathology Beaumont Hospital/RCSI Medical School Dublin

Context

Chronic hyponatremia is a rare clinical entity, encountered predominantly in the elderly population.

In the younger population, chronic hyponatremia is often a consequence of failure to generate thirst in response to osmotic stimuli.

We report the first case of a patient with a disconnect between normal osmoregulated thirst and abnormal drinking behaviour.

Case report

A 24 year old man presented with gait instability, myalgia, and cognitive decline. He was dehydrated and had marked facial dysmorphism. No focal neurological signs were noted on examination. The patient's biochemical picture was consistent with hyponatremic dehydration (Table 1).

Table 1. Results of laboratory investigations during admission

Parameter	Day 1	Day 2	Day 3	Day 4	Day 5
Na (ref 133-146 mmol/L)	175	170	161	150	140
K (ref 3.5-5.3 mmol/L)	2.2	2.3	4.1	4.2	3.9
Urea (ref 2.7-7.8 mmol/L)	16.9	15.6	12.7	11.3	8
Creatinine (ref 64-104 mmol/L)	135	128	108	90	76
Posm (ref 275-295 mosm/kg)	374	-	344	-	296
Uosm	894	963	-	-	496
CK (ref 0-170 U/L)	15540	41297	37260	5288	2320

MRI brain showed marked hydrocephalus (Image 1).

The patient was treated with IV dextrose and recovered without neurologic sequelae.

A reset osmostat for thirst and AVP release was suspected and following discharge, the patient underwent 5% saline infusion.¹ Plasma AVP rose from 1.4 to 7.3 pmol/l and linear regression analysis defined a normal osmotic threshold for AVP release of 283 mOsm/kg; $pAVP = 0.27 (pOsm - 283)$, $r = 0.88$, $p = 0.002$. Thirst (visual analogue scale) rose appropriately, with a normal osmotic threshold: $thirst = 0.31 (pOsm - 283)$, $r = 0.98$, $p < 0.0001$. The patient therefore had normal osmoregulatory function (Fig 1).

However, in the 30 mins following infusion, the patient only drank 400 mls water, despite normal thirst dynamics (normal water intake 700-1200 mls).¹

Therefore, there was a disconnect between normal osmoregulated thirst and his abnormal drinking behaviour.

The patient was recommended to have a fixed fluid intake of 2-3 litres daily. He remained eunatremic until ten years later, when during febrile illness, his fluid intake fell and he again presented with hyponatremic dehydration.

The patient has had no progression in his neuroimaging and no development of neurological features after 10 years of follow-up.

Image 1. MRI brain showing large extra-axial cyst with mass effect and complete absence of corpus callosum

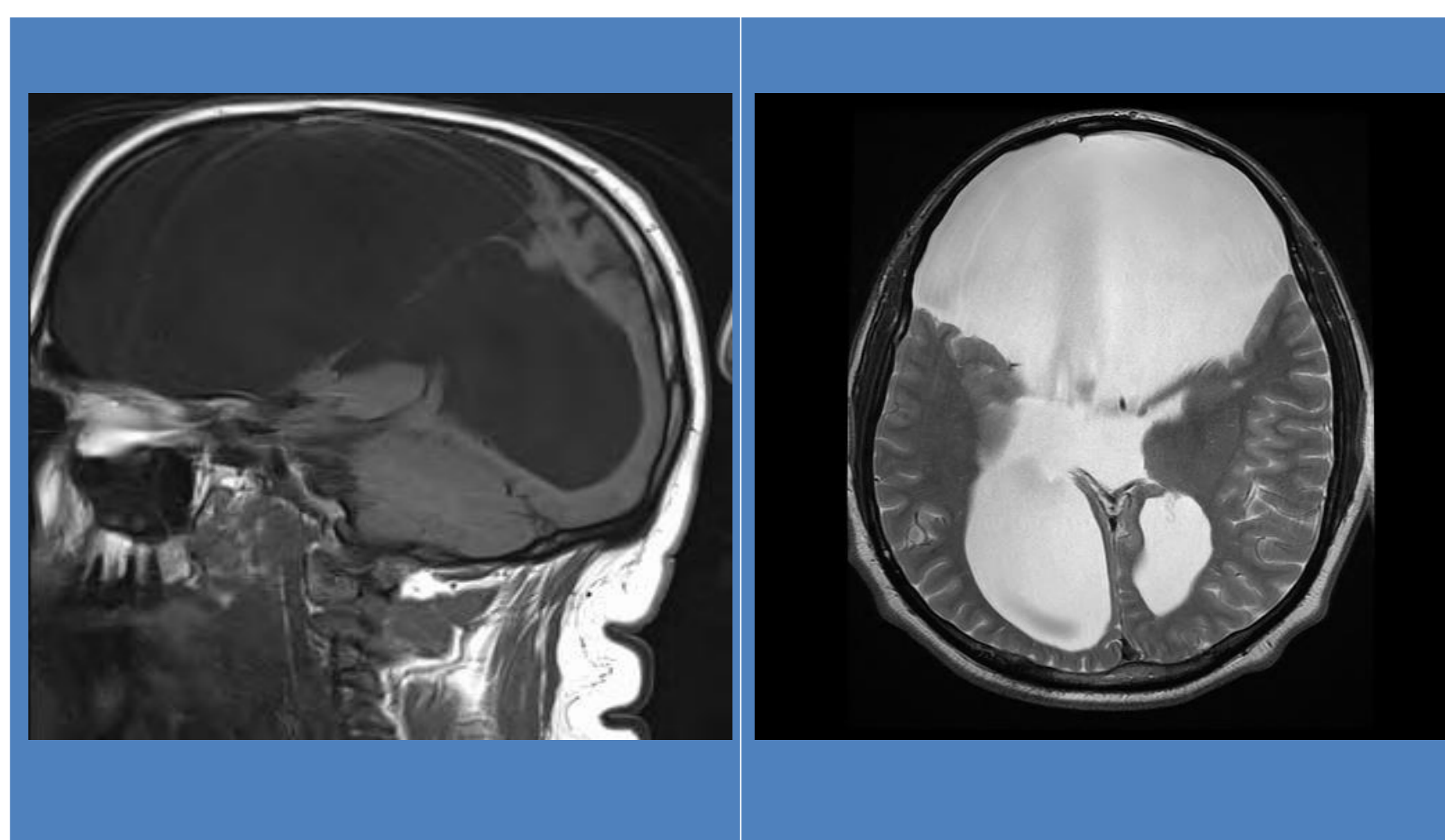
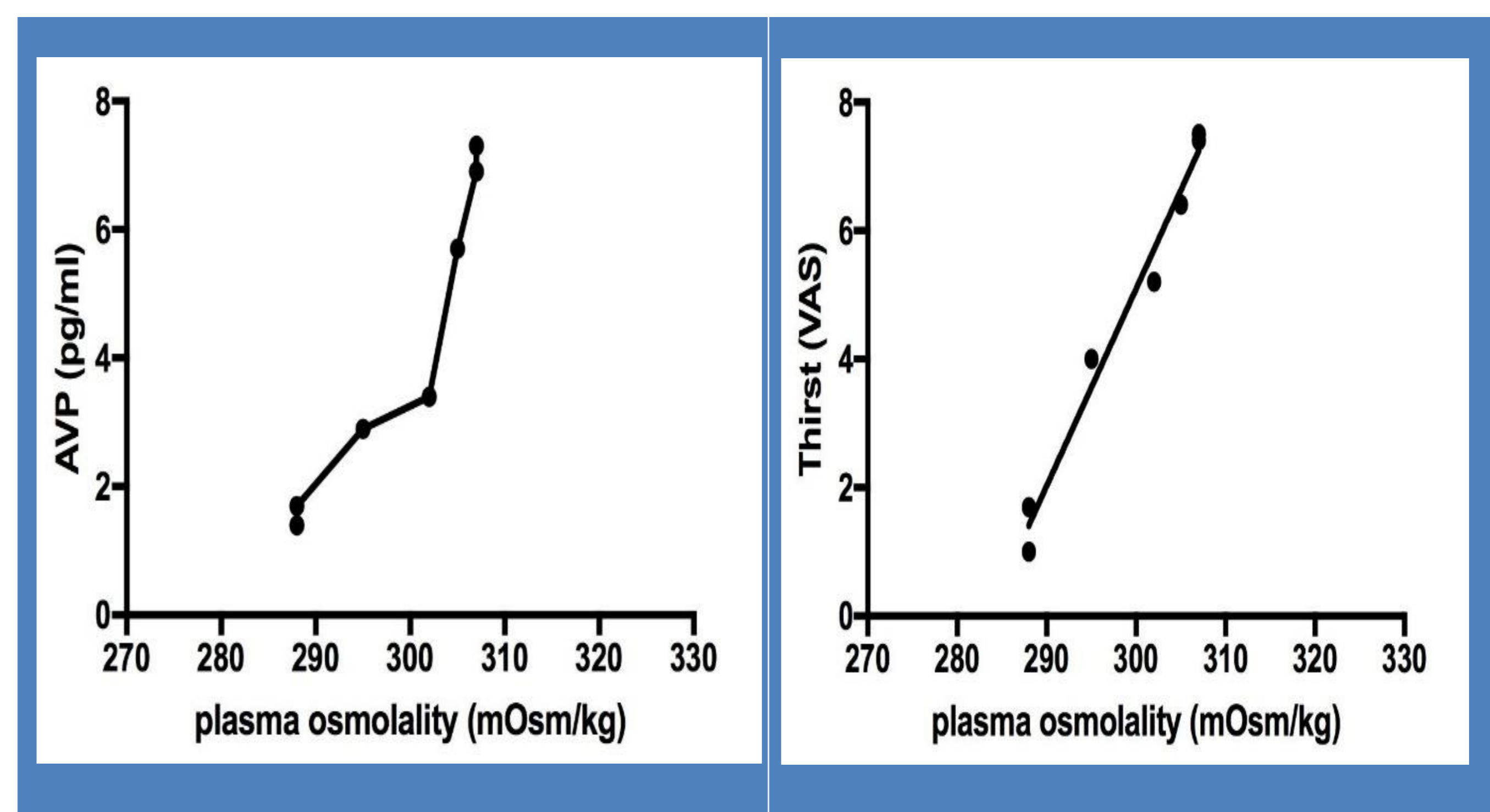


Fig 1. Schematic representation of plasma vasopressin release and thirst after infusion of hypertonic saline in our patient. The response is within normal range.¹



Key points

- Our patient has an unique disconnect between the osmoregulatory function and fluid intake.
- The mechanism of this abnormality is unknown.
- It is difficult to dissociate this patient's abnormal drinking behaviour from his marked hydrocephalus, as distortion of the complex anatomic structures that translate thirst appreciation into the central drive to drink seems inevitable.³
- Learned behaviour with fixed fluid intake has prevented day today hyponatremia, but the patient remains vulnerable to severe life threatening hyponatremia during intercurrent febrile illnesses.

References

1. Thompson CJ, Bland J, Burd J et al.: The osmotic thresholds for thirst and vasopressin release are similar in healthy man. *Clin Sci (Lond)* 1986; 71(6):651-6.
2. Baylis PH, Thompson CJ: Osmoregulation of vasopressin secretion and thirst in health and disease. *Clin Endocrinol.* 1988; 29:549-576.
3. Egan G, Silk T, Zamarippa F, et al: Neural correlates of the emergence of consciousness of thirst. *Proc Natl Acad Sci U S A.* 2003; 100(25):15241-6.