Description of Patientes Diagnosed with Central Diabetes Insipidus, 14 years experience at the National´s Children Hospital, Costa Rica

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ABSTRACT

Objective and hypotheses

To describe the epidemiology of patients diagnosed with central diabetes insipidus (CDI) in a National Children's Hospital, from January 2000 to December 2013. Is thought to be a disease with a low incidence in our country.

Methodology

A retrospective study based on the clinical records of patients with CDI. Were analysed a total of 30 patients, after exclusion of some records. Qualitative and quantitative analysis of variables, measures of central tendency were used (mean, median and frequencies).

Results

Of the total of 30 patients, there was a sex distribuion of 17 males and 13 females, no statistical difference. The incidence of central diabetes insipidus was of 2.6 cases per 100 000 habitants. The mean age at presentation was 60 months (1–154 months). At diagnosis, 83% of patients reported polyuria and was documented hypernatremia in 100% of cases and hyperosmolarity in 83%. The most common cause of CDI was the inmediate posoperative period of suprasellar tumors, in 11 of the 30 patients (36%); which the most frequent was craniopharyngioma (64% of the tumors and 50% of the total CDI). The other most common hormone deficiency added to the CDI was central hypothyroidism, in a total of 11 patients.

Conclusion

CDI in Costa Rica is a rare problem occurring in childhood at a mean age of 5 years. The most common cause are suprasellar tumors, of which the most frequent is the craniopharyngioma. Follow up of patients without an etiology of CDI is crucial.

BACKGROUND

Diabetes insipidus (DI) is a heterogeneous clinical syndrome involving an alteration in water balance. When caused by a deficiency in the production of antidiuretic hormone (ADH) it's called central diabetes insipidus (CDI).

DI patients typically present with polyuria, nicturia, and polydipsia due to the initial rise in serum sodium and plasma osmolarity. They can also present with severe dehydration, vomiting, constipation, fever, irritability, altered sleep pattern and fail to thrive.

There are few prospective studies about the etiology of CDI in children, and virtually in all retrospective studies, many of the causes are determined as idiopathic (10.4–55.2%, according to various authors). In a recent prospective study, in 85 patients from a single tertiary care center was possible to establish the etiology of CDI up to 96% of them after a period of radioimaging and monitoring the Pituitary-thyroid-adrenal-gonadal axis for at least 5 years.

There are no previous studies in our population.

METHODS

A retrospective study based on the clinical records of patients with central diabetes insipidus. Records were obtained from January 2000 to December 2013. Were analysed a total of 30 patients, after exclusion of some records. Qualitative and quantitative analysis of variables, measures of central tendency were used (mean, median and frequencies), using Epi Info v7 y Excel 2010.

RESULTS

Of the total of 30 patients, there was a sex distribuion of 17 males and 13 females, no statistical difference. The incidence of central diabetes insipidus was of 2.6 cases per 100 000 habitants. The mean age at presentation was 60 months (1–154 months).

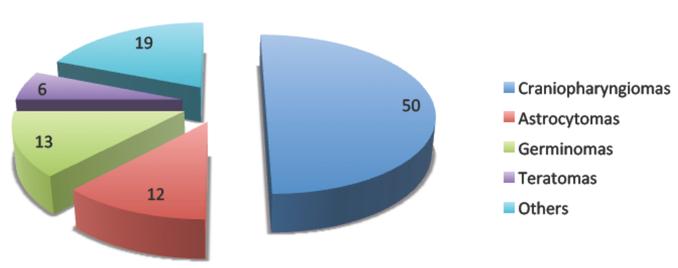
At diagnosis, 83% of patients reported polyuria and 43% polydipsia. The average urine output was 8cc per kilogram per hour (range of 5-24). It was found that the vast majority of children were eutrophic (46.7%), 7 of the 30 obese or overweight and 5 of them with malnutrition; short stature in 24% of the cases. When examining the causes of CDI, in 4 patients (13.3%) initially the diagnosis was not done; only 1 was reported later with thickening of pituitary stalk . The most common cause of CDI were tumors of the CNS, whether the diagnosis was made after the neurosurgical intervention (11, 36.7% of the total), or as a result of the approach to patients who develop CDI (5, 16.7%) (Table 1). At routine laboratories, hypernatremia was documented in 100% of cases and hyperosmolarity in 83%.

Table 1. Clinical presentation and etiology of CDI (n:30)

Signs and Symptoms	n	%
Polyuria	25	83,3
Polydipsia	13	43,3
Nutritional status	**n**	**%**
Normal	14	46,7
Overweight-obesity	7	23,3
Failure to thrive-desnutrition	5	16,7
Not reported	4	13,3
Etiology	**n**	**%**
Neurosurgery posoperatory	11	36,7
CNS tumor diagnosed after CDI	5	16,7
Central midline defect	4	13,3
Histiocytosis	3	10,0
CNS infection	2	6,7
Head trauma	1	3,3
Not diagnosed	4	13,3
TOTAL	30	100

CNS tumors in patients to whom neurosurgery was performed, and as a result developed CDI, 64% correspond to Craniopharyngiomas. Analyzing the total of patients who had a CNS tumor, 50% of the cases were craniopharyngiomas. Other tumors observed: astrocytomas, germinomas and teratomas (Figure 1).

Figure 1: Tumors associated to CDI (%)



Studies were done to rule out involvement of other pituitary axis hormones. In eleven of the thirty patients were compromised thyroid axis hormones, growth hormone (GH) and/or ACTH-cortisol. In 100% of cases it was established central deficiency of thyroid hormone, in 9 of 11 patients was documented ACTH-cortisol deficiency, and in only 1 patient was established the diagnosis of GH deficiency. (Table 2)

Table 2. Other pituitary deficiencies (n:30)

	n	%
Absent	15	50.0
Not studied	4	13.3
Present	11	36.7
Thyroid	11	100
ACTH-cortisol	9	81.0
GH	1	9.0
TOTAL	30	100

DISCUSION

The incidence of CDI is variable and depends on the underlying cause. In this study it was possible to demonstrate an incidence of 2.6 cases per 100 000 habitants, similar to that reported by Juul et al in Denmark, which found that the annual incidence of CDI was 3-4 per 100 000 habitants. The mean age at presentation was 5 years; 7.5 years is described in other places, concluding that in our country we make early diagnosis and/or the appearance of certain tumors occurs at a younger age. As in all studies, polyuria is the cardinal symptom of the disease.

The percentage of idiopathic CDI in the literature varies from 10 to 50%, which is consistent with our study (13%). When the CDI is associated with craniopharyngioma, it usually involves other adenohypophysis hormones. In our analysis 100% had hypothyroidism and only one case of growth hormone deficiency, which is not consistent with reports worldwide and we should study further.

Monitoring of patients with idiopathic CDI is important because there are reported cases of diseases late appearance, especially histiocytosis, even up to 10 years after the onset of CDI. In our series of 4 patients without a diagnosis, only in one a thickening of the pituitary stalk was observed later without a clear diagnosis.

Limitation of retrospective research is its dependence on medical record documentation quality. We believe that this factor has minimal impact on the current study, because the information we abstracted from records was objective for the majority of the variables.

CONCLUSIONS

Central diabetes insipidus in Costa Rica is a rare problem occurring in childhood at a mean age of 5 years. The most common cause are suprasellar tumors, of which the most frequent is the craniopharyngioma. Follow up of patients without an etiology of CDI is crucial.

BIBLIOGRAPHY

1. Di Iorgi Natascia et al. Diabetes Insipidus – Diagnosis and Management. Horm Res Paediatr, 2012.
2. Argente, Jesús. Diabetes Insípida. Editorial Justim S.L., 2009.
3. Di Iorgi Natascia. Central Diabetes Insipidus in Children and Young Adults: Etiological Diagnosis and Long - Term Outcome of Idiopathic Cases. J Clin Endocrinol Metab, 2013.
4. Juul KV et al. National Surveillance of Central Diabetes Insipidus (CDI) in Denmark: Results from 5-years Registration of 9,309 Prescriptions of Desmopressin to 1,285 CDI patients. J Clin Endocrinol Metab, 2014.
5. Ali Varan. Evaluation of Patients with Intracranial Tumors and Central Diabetes Insipidus. Pediatric Hematology and Oncology, 2013.

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