



Pediatric Diabetes Insipidus



Irina Suley Tirado Pérez^{1*} and Andrea Carolina Zárate Vergara²

¹Medical Epidemiologist, Master Pediatric Palliative Care, University of Santander, Colombia

²Medical Epidemiologist, Graduate Student Pediatric Intensive Care, University of Santander, Colombia

*Corresponding author: Irina Suley Tirado Pérez, Medical Epidemiologist, Master Pediatric Palliative Care, Graduate Student Pediatric Intensive Care, University of Santander, Colombia

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Abstract

Diabetes insipidus is a rare condition, with classic signs of clinical presentation and are polyuria and polydipsia, presented by excessive fluid intake, absence or resistance to the action of antidiuretic hormone, below is a topic review will be made explaining the pathophysiological characteristics of this disease, its diagnosis and treatment.

Keywords: Diabetes insipidus; Polyuria; Polydipsia; Vasopressin

Introduction

Mainly diabetes insipidus is characterized by clinical symptoms such as polyuria and polydipsia, secondary to absolute or relative lack of secretion or action of antidiuretic hormone (ADH), also called vasopressin, three presentations, primary polydipsia when much ingests liquid, which suppresses ADH, the other presentations is called central diabetes insipidus when there is deficiency of this hormone, as nephrogenic diabetes insipidus is due to a resistance to the action of vasopressin, these entities may arise from hereditary defects or other causes [1,2].

Its cause is usually due to destruction or degeneration of neurons originate in the supraoptic and paraventricular nuclei. Other causes include inflammatory and local autoimmune diseases, vascular diseases, intracranial tumors, trauma or brain malformations, histiocytosis Langerhans, sarcoidosis, other genetic causes, is genetic defects in the synthesis of arginine vasopressin (AVP) inherited as autosomal dominant, autosomal recessive, or X-linked recessive traits, nephrogenic diabetes insipidus secondary to mutations in the AVP receptor V2 (AVPR2), resulting in a loss of function or dysregulation of AVPR2 renal, also described as causal abnormalities aquaporin 2 (AQP2) water channel gene [3,4].

Definitions

Dipsogénica diabetes insipidus or primary polydipsia

Primary polydipsia occurs because the amount of liquid ingested by the patient is much higher than the requirement hypersensitivity center thirst, thereby decreasing the osmolality of the internal environment, causing a suppression of secretion of ADH, thus giving the clinical feature is polyuria, occurs in patients

with psychiatric, anxious or diseases drugs such as phenothiazines [1,5,6].

Central diabetes insipidus

Central diabetes insipidus caused by deficiency of arginine vasopressin, also called cranial diabetes insipidus or hypothalamic, may have a sporadic or familial behavior, central diabetes insipidus sporadic may be due to causes that alter the hypothalamus or hypothalamohypophysial area, as craniopharyngioma, may present clinically isolated or associated with deficits of anterior pituitary hormones; central diabetes insipidus family is presented by mutations in the coding for ADH, which is located chromosome 20p13 in with an autosomal dominant pattern of inheritance, clinical presentation may be from infancy to adult [3,5]. In a retrospective study, conducted in Peru in pediatric population was observed that the most common cause of central diabetes insipidus intracranial tumors were at 55.7%, then of Langerhans Cell Histiocytosis with 13.9% and malformations brain with 8.9% and 17.7% idiopathic causes [7,8].

Nephrogenic diabetes insipidus

Nephrogenic diabetes insipidus is rare, it is presented by the inability to concentrate urine, although their antidiuretic hormone, occurs most X-linked, recessively frequency, and is seen in disorder affecting renal tubular function, either urologic obstructive disease, renal failure, renal cystic disease, interstitial nephritis, among others, and can be sporadic and family. Sporadic mainly occurs by hyperglycemia, which causes osmotic diuresis, inverting the gradient across the renal tubule, diabetes insipidus family is linked

to the X chromosome, specifically there alteration in Xq28, and its most severe clinical presentation it occurs during lactation [5,9,10].

Epidemiology

Diabetes insipidus is a rare entity, it is estimated in the literature that has a prevalence of 1 per 25000 inhabitants, being most often central diabetes insipidus, which nephrogenic diabetes insipidus, with a smaller percentage etiology of hereditary nature less than 10%; diabetes insipidus in the raw X-linked, reporting 4 to 8 cases per million live births [3,11].

Pathophysiology

Water homeostasis is balanced by release of the antidiuretic hormone vasopressin and stimulation of the thirst. Vasopressin is produced in the magnocellular neurons of the paraventricular and supraoptic nuclei of the hypothalamus that send axons to the posterior pituitary nuclei, which is responsible for segregating arginine vasopressin into the bloodstream where it acts in the kidney duct renal collecting increasing the reabsorption water duct renal collecting (eleven).

Vasopressin is secreted response fluid volume changes, acting as principal regulator of water permeability in the renal collecting ducts, aquaporin are essential water permeability of the apical membranes mainly AQP2, in the absence of signaling AVP, no such permeability. When binding AVP receptor V2 AVPR2 exists, a signaling cascade ultimately leading starts insertion AQP2 channels in the apical membrane AVPR2, this binding leads to the release of stimulatory G protein being a receptor bound to this protein, which in turn activates adenylyl cyclase and simultaneously increases the levels of cAMP activates protein kinase a (PKA), there mediated phosphorylation of AQP2 PKA, stimulation of AQP2 potentiates allowing water permeability and water reabsorption in the collecting system [12-14].

The release and synthesis of AVP is primarily regulated by plasma osmolality gap or eletrolitros as sodium, while the latter increase produces a significantly stimulate release and transcription AVP gene, When there is a deficiency of AVP, known as central diabetes insipidus, leading to hypotonic polyuria, when there is resistance arginine vasopressin renal distal tubule, leading to a deficiency in tubular reabsorption of water occurs nephrogenic diabetes insipidus [8,13].

Diagnosis

Diagnosis requires a complete medical history and physical examination, the main symptoms are polyuria and polydipsia, many children have decreased weight excluding the fluid, or in children greater intellectual involvement by pictures of repetitive dehydration, these episodes of dehydration when severe can lead to seizures. It can also present vomiting, constipation, fever, irritability, sleep disorders, growth retardation and stunting, when having visual impairment should be thought of brain tumors or associated symptoms with other diseases such as septo-optic

dysplasia consisting defects midline, nystagmus, hypoglycemia, jaundice and micropenis [15,16].

Known as polyuria urine production 24 hours, with a fluid intake than 2l/m²/24h or about 150mL/kg/ 24hr at birth, 100-110mL/kg/24h up to the age of 2 years and 40-50mL/kg/24h in older children and adults, should apply eletrolitos serum as potassium, calcium, glucose and creatinine for differential diagnosis, is also very useful measure urine osmolality <300mOsm/kg and Osm>300mOsm/kg serum think diabetes insipidus, while discarded if urine Osm>600mOsm/kg, or serum Osm<270mOsm/kg, and if there symptomatology should be performed deprivation test water [2,16].

Images are requirement to assess the etiology of diabetes insipidus, especially MRI to look for brain tumors, this posterior pituitary can be seen as a hyperintense on sagittal T1 images, the lack of this in the posterior pituitary can mean disorders and identify at an early age of local tumors hidden [3,17].

Treatment

The main objective of management for diabetes insipidus is decreased urine output and thirst control, allowing adequate control of levels of sodium and water homeostasis, the management of primary forms of diabetes insipidus focuses on dietary modification to reduce the salt load. In central diabetes insipidus the fluid therapy plays an important role in newborns and infants require large volumes (3L/m²/24h), accompanied by analogue of vasopressin as desmopressin have a good result and prevents complications, is in various presentations as oral, intranasal, or subcutaneous preparation, oral is preferred over intranasal, and on subcutaneous for ease in administration, oral dose is from 0.025 to 1.2mg/day in 1-3 doses, 5-10mcg intranasal, 0.1-0.2mcg subcutaneous or continuous infusion to vasopressin 3.1mU/kg/hr, in an intensive care unit [2,8,11].

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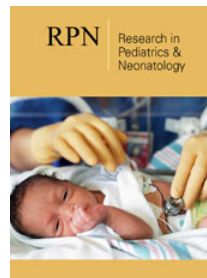
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