

Aetiology and different clinical conditions of adrenal insufficiency in a region of North Africa

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INTRODUCTION

Adrenal insufficiency is relatively rare in childhood and adolescence. Signs and symptoms may be non specific; therefore, the diagnosis may not be suspected early in the course. It may be categorized as primary or secondary and congenital or acquired. Primary adrenal insufficiency (PAI) is uncommon in the Western population and is estimated to affect 90 to 140 per 1million people. Many etiologies have been reported worldwide of which CAH was the commonest etiology in children.

AIM

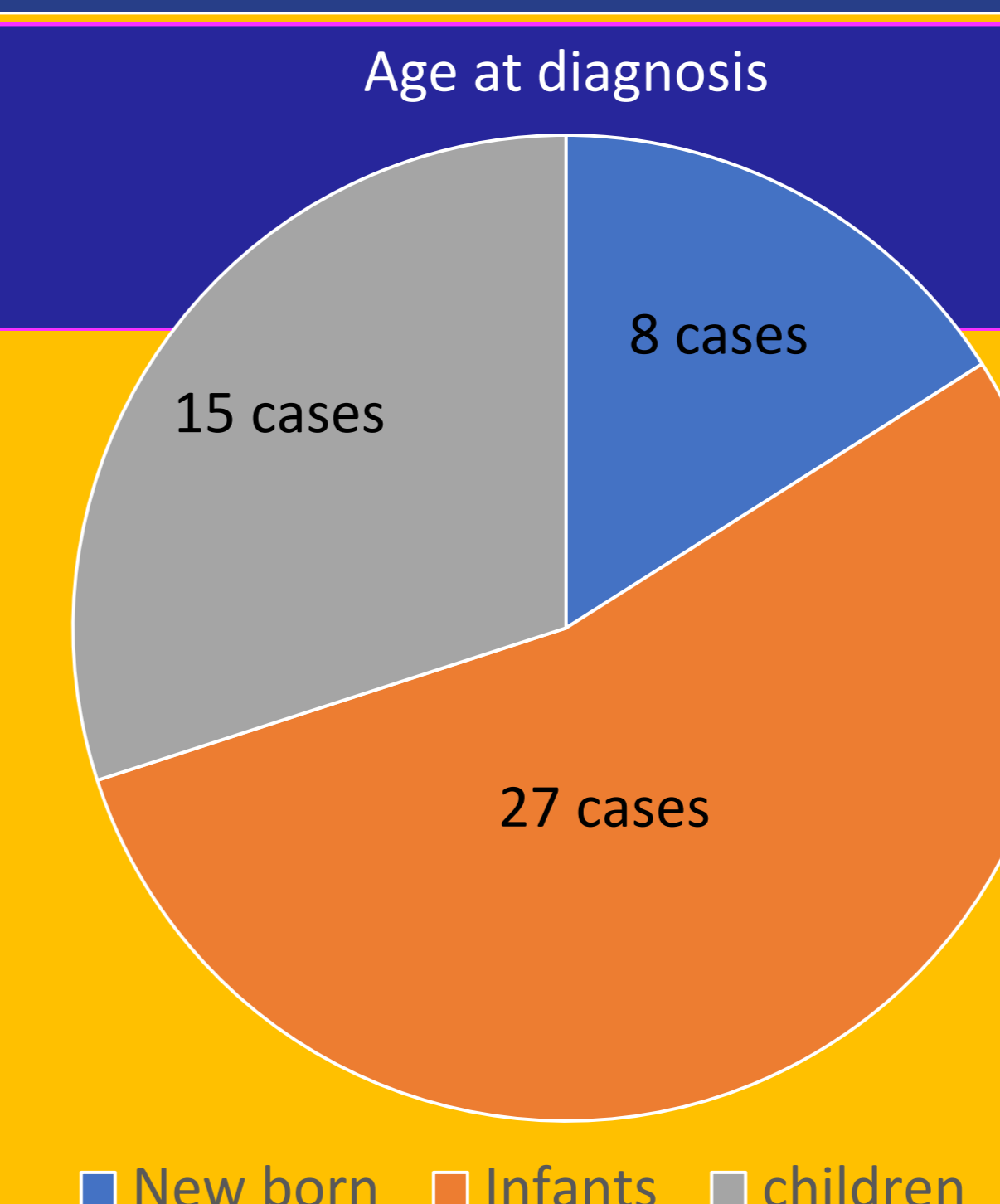
To determine the clinical features and evolution of childhood primary adrenal insufficiency in our centre.

MÉTHOD

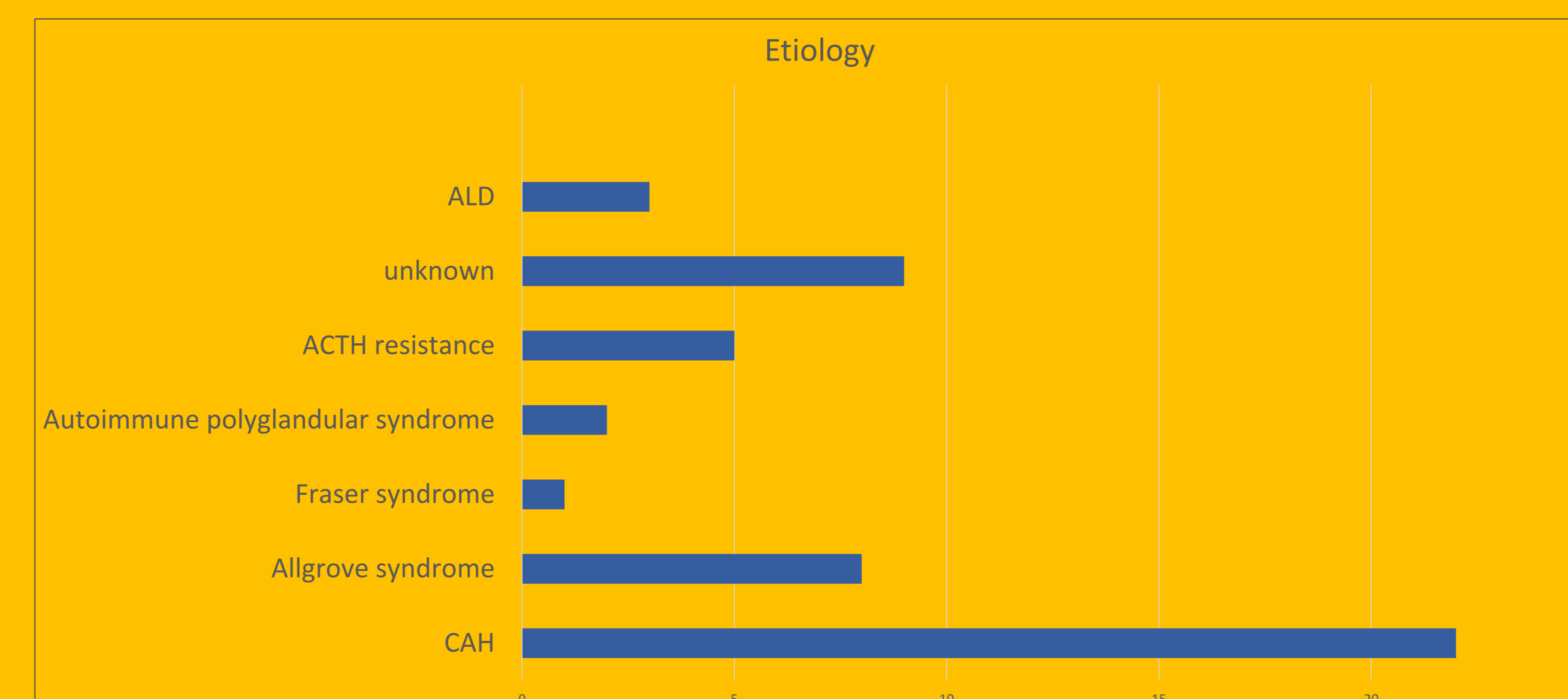
Longitudinal retrospective study of patients diagnosed with PAI from January 2005 to December 2019. Clinical features at presentation as well as the possible etiology were described, investigations and management.

RESULTS

Between 2005 and 2019, 50 patients (36 girls, 14 boys) were followed for a mean (range) duration of 7.5 [1-14] years. Mean age at diagnosis and start of treatment was 16.3[0.4-60] months. Consanguinity was present in 28(56%) of cases, and there were 7 familial cases (14%). Of 22 cases presented CAH (44 %), 14 (64 %) had virilized genitalia (females), 4 boys were diagnosed after salt wasting crisis, 4 cases had presented signs of androgen excess. Of the 28 cases presented other etiologies, symptoms at presentation included melanoderma, fatigability, abdominal pain, diarrhea vomiting and shock. Delay in diagnosis was observed in 13 cases (46 %). Aetiology was unknown in 9 cases who need a genetic investigation, The most common etiology was Allgrove syndrome (8cases). Mean ACTH at diagnosis was 1746 ± 1731 pg/ml. ACTH test have been realized in 8 cases to confirm a diagnosis. 17 OHP was high in 20 cases with CAH, abdomen ultrasononography was realized in 32 cases. Antibodies investigation is not available in our country.



Symptoms	Number	Frequency %
fatigability	38	76
Abdominal pain	3	6
Melanoderma	13	26
Dehydration	18	36
Alacrymia	8	16
Dysphagia	3	6
Vomiting	14	28
Shock	2	4
DSD	20	40
Adrenal crisis	37	74



COMMENTS

Adrenal disorders can be difficult to diagnose and are associated with considerable morbidity and mortality if undiagnosed. The clinical and biochemical features at presentation and the natural history of the condition vary depending on which adrenal steroids are affected, as well as the underlying pathological condition. Although a specific diagnosis can be reached in many children and young people with PAI, sometimes the cause remains unknown. Our experience underlines the need to establish links with centres which are able to carry out the necessary genetic analyses in our patients.

CONCLUSION

Adrenal insufficiency is rare, but fatal medical condition. The most common causes is CAH. Causes other than CAH should be suspected during evaluation of PAI. Many patients remained with unknown etiologic diagnosis, and need more specific tests and genetic that are not available in our country, hence the need for collaboration with specialized centers

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