INTRODUCTION

Allgrove syndrome is a rare autosomal recessive disorder involving alacrymia, achalasia, Addison's disease (3A) and neurological disorders (4A), it results from mutations in the AAAS gene located on chromosome 12q13 which codes for a protein known as ALADIN (Alacrymia Achalasia aDrenal Insufficiency Neurologic disorder).
Alacrymia is diagnosed by Schirmer's test, achalasia by esophageal manometry while adrenal insufficiency is confirmed by the determination of cortisol and ACTH.

AIM

To describe the clinical and evolutionary aspects of patients who presented Allgrove syndrome in our department.

MÉTHOD

It is a retrospective, descriptive study, spanning the period 2010 to 2020; were eligible for the study all patients who were hospitalized for Allgrove syndrome. Several parameters were evaluated, age, sex, diagnostic elements, dose of treatment and evolution.

RÉSULTS

Age at diagnosis: 5 years
diagnostic deadline: 3 years.
Sexe ratio: 3 girls for 1 boy.
Consanguinity in 1 patient (syndrome Allgrove family).
Age of the first symptoms: 2 years.
Alacrymia confirmed by the test of Schirmer
Achalasia confirmed by endoscopy esogastro-duodenal and TOGD.
All the patients were treated by glucocorticoids, tears artificial.
On average, 5 dilation sessions were performed [2-11].

Evolution: We have an average follow-up of 4 years
(6 months - 8 years).
Improvement in adrenal insufficiency was achieved in all patients.
Successful dilations were obtained in 2 cases, the other 2 underwent a successful Heller cardiomyotomy.

REFERENCES