European Cystic Fibrosis Society

Journal of Cystic Fibrosis
The Official Journal of the European Cystic Fibrosis Society

# Cystic Fibrosis Research News

## Title:

Characteristics of Electrolyte Imbalance and Pseudo-Bartter Syndrome in Hospitalized Cystic Fibrosis Children and Adolescents.

## Lay Title:

Features of Salt Imbalance and Pseudo Barter Syndrome in hospitalized Children and Young Adults with Cystic Fibrosis.

## Authors:

Danish Abdul Aziz (MBBS, MRCPCH (UK), FCPS)[1] Fatima Siddiqui (MBBS)[2] Quratulain Abbasi (MBBS) [3] Haissan Iftikhar(MBBS,FCPS) [4] Shahira Shahid(MBBS) [5] Fatima Mir(MBBS,FCPS)[6]

## Affiliations:

1. Assistant Professor, Department of Paediatrics and Child Health, Aga Khan University Hospital Karachi, Pakistan.
2. Intern, Dr. Ruth K.M Pfau Civil Hospital Karachi, Pakistan.
3. Resident Paediatrics, Department of Paediatrics and Child Health, Aga Khan University Hospital Karachi, Pakistan.
4. Fellow ENT, Department of otorhinolaryngology, Aga Khan University Hospital Karachi, Pakistan.
5. Research Assistant, Department of Paediatrics and Child Health, Aga Khan University Hospital Karachi, Pakistan.
6. Associate Professor, Department of Paediatrics and Child Health, Aga Khan University Hospital Karachi, Pakistan.

## What was your research question?

To find out if there are differences in the clinical features between young people with Cystic Fibrosis (CF) who present with either 1) salt imbalance, 2) Pseudo-bartter syndrome (PBS) or 3) normal salt level in blood.

## Why is this important?

Pseudo-bartter syndrome (PBS) is an unusual yet an important presentation in people with CF. This syndrome can present with abnormal blood salt levels (low sodium, low potassium, low chloride and increased bicarbonate ion concentration) and higher blood pH but with normal salt excretion in urine. Babies can even present with PBS before their diagnosis of CF.

# Cystic Fibrosis Research News

Therefore it is important to suspect PBS in patients who may be undergoing cystic fibrosis diagnosis

## What did you do?

In our study, we included people with a positive genetic test for CF, increased sweat chloride levels, clinical features of lung disease, positive family history, or pancreatic insufficiency in the form of long-standing pasty stool.
These people were then divided into three groups, 1) salt imbalance, without complete features of PBS, 2) diagnosis of PBS, and 3) normal salt level.
These three groups were studied for their age, gender, weight, height, number of lung infections, days spent in hospital, and if they acquired Pseudomonas aeruginosa. The results of these outcomes were than compared between the three groups.

## What did you find?

26% of the people studied had electrolyte imbalance on presentation while 15% had PBS. Of those patients who presented with PBS, half (7.5%) had PBS as their initial presentation to medical care which then led to the diagnosis of CF. This half were termed as 'early presenters'. The other half, who were first diagnosed with CF and later presented with PBS, were termed as 'diagnosed CF patients'. Early presenters had no history of lung problems and increased urinary frequency was the most common symptom at presentation in them. Infections with Pseudomonas aeruginosa were more common in diagnosed CF patients.

## What does this mean and reasons for caution?

People with CF can present with PBS anytime during the course of their illness. Therefore, any patient presenting with CF should be checked and treated for salt imbalance. Additionally, in patients presenting with features of PBS having dehydration or laboratory evidence of salt imbalance peculiar to PBS, with no previous history of lung related disease, should be carefully assessed for CF. The limitation of our study was that it was a document review of clinical features of already diagnosed CF patients and their laboratory findings.

## What's next?

Further large multi-centre prospective studies are needed that include patients presenting with suspicion of abnormal salt levels and dehydration to help in improving their quality of life through timely diagnosis of CF and early intervention.

# Cystic Fibrosis Research News

**Original manuscript citation in PubMed**
https://pubmed.ncbi.nlm.nih.gov/34610890/