

Location: Office

Presenting complaint: A 2-year-old boy is brought with complaints of failure to gain weight and loose stools.

Vitals: Pulse: 100/min, B.P: 80/56 mm Hg, Temp: 101.2 F, R.R: 18/min, Height: 27.2 inches (68cm), Weight: 7.2 Kg (16 lbs)

HPI:

A 2-year-old Caucasian child is brought to the doctor's office by his parents for an evaluation of loose greasy stools and failure to gain weight despite adequate nutrition. His other problems are intermittent productive cough and rhinorrhea. The parents state that he has been wheezing and coughing up a purulent expectoration for 4 days. They deny fever or chills. He has had pneumonia 4 times since birth. Delivery of the child and neonatal course were uncomplicated. He was breast-fed until the age of 4 months. There is a history of CF in the family of both of his parents.

Review of Systems:

General:

Skin: No rashes or lesions

HEENT: Nasal discharge

Musculoskeletal: No joint swelling

Cardio respiratory: see HPI

Genitourinary: No complaints

Abdominal: see HPI

Development: Delayed

Vaccinations: Up-to-date

How to approach this case:

This child presents with failure to thrive. Failure to thrive has both organic and non-organic causes. The etiology of failure to thrive in this child is most likely cystic fibrosis. Clues to cystic fibrosis in this patient are positive family history, repeated chest infections and malabsorptive diarrhea.

Order:

Complete physical examination (except breast)

RESULTS:

The child seems to be emaciated; his height and weight are lower than expected for his age. Chest examination shows generalized hyper-resonance and scattered crepitations bilaterally. He also has wheezing especially during expiration.

ORDER REVIEW:

Admit to the ward

Pulse oximetry, stat and every 4 hours

IV access, stat

Sputum gram stain, stat

Sputum culture and sensitivity, stat (Type sputum C&S)

Blood cultures, stat

CBC with differential, stat

BMP, stat

CXR-PA/Lateral, stat

Sinus x-ray, routine

Sweat chloride, routine

72-hour fecal fat estimation, routine

Treatment:

Oxygen inhalation, continuous (If saturation are <92% on room air)

Amoxicillin and Clavulonic acid, oral, continuous (Type Augmentin)
Nebulized Albuterol, inhalation, continuous (4 times a day)
Multivitamin tablets, oral, continuous
Chest physiotherapy
Vitals Q 6 hours
D5NS, IV, continuous
Regular diet
Ambulation at will

RESULTS:

CBC shows neutrophilic leukocytosis
Sweat chloride is 85 meq/L
BMP showed low sodium and potassium
CXR shows hyperinflation of both lung fields.
X-ray PNS shows opacification of paranasal sinuses.
Gram staining of sputum does not show any predominant organism.
Sputum culture is pending.
72-hr fecal fat estimation is pending

DISCUSSION:

Cystic fibrosis is an autosomal recessive disorder commonly affecting Caucasians. Its clinical manifestations include acute or persistent respiratory symptoms, failure to thrive, meconium ileus, diarrhea, rectal prolapse, nasal polyps, electrolyte or acid-base disorders and hepatobiliary disease. Diagnosis of cystic fibrosis is made when evidence of CFTR dysfunction is present along with typical clinical features or positive family history. Elevated sweat chloride on two separate occasions is an evidence of CFTR dysfunction. Other important tests are gram stain, culture and sensitivity of the sputum, chest X-ray, X-ray of the para nasal sinuses. 24 hour fecal fat estimation should be used to diagnose the malabsorption. Genetic testing is not required to make the diagnosis as it is associated with multiple mutations.

All children who are suspected to be suffering from CF should be admitted to the hospital. Evaluation should include baseline testing, accurate diagnosis, initiation of treatment and education of the patient and parents. Follow-up should be done every 2-3 months for monitoring. History, physical examination and staining and culture of sputum or pharyngeal swab are required on each follow-up visit. Prophylactic immunization against influenza, measles, and pertussis should be given.

Treatment:

- Antibiotics are given when a patient of CF develops acute or sub-acute increase in sputum production, cough, dyspnea and/or fever. Antibiotics are selected according to the results of sputum culture. Sputum cultures are performed at least on yearly basis to help identify the bacteria, which are chronically inhabiting the respiratory tract. Oral antibiotics are used when exacerbation is mild, while IV antibiotics are used when exacerbation is severe and when bacteria are resistant to oral antibiotics. For *S. aureus*, oral antibiotics used are cephalexin, dicloxacillin or amoxicillin-clavulanate. For *P. aeruginosa*, ciprofloxacin is used as an oral antibiotic. IV regimen for *P. aeruginosa* is a combination of tobramycin and anti-pseudomonal penicillin like piperacillin.
- Bronchodilators like albuterol or salmeterol are used in patients with airflow obstruction.
- DNase is usually prescribed for those with daily productive cough plus airflow obstruction.
- Combination of physiotherapy and exercise should be considered in patients with more retained purulent secretions.
- Inhaled glucocorticoids can be used in patients of CF who have clinical evidence of airway hyperactivity.
- Oxygen therapy should be considered in all patients who have evidence of hypoxemia at night or rest or pulmonary HTN.

- Lung transplantation is the only definitive treatment in patients with severe infections and grossly damaged lungs with a FEV1 of 30% or less of the predicted value.
- Nutrition: Recommended diet is one with high protein and high calories. Vitamin supplementation of fat-soluble vitamins and pancreatic enzyme replacement are also very important.

Results review:

72-hours fecal fat is elevated.

Sputum culture grew staphylococcus aureus, sensitive to cephalexin.

ORDER REVIEW:

D/C Amoxicillin and Clavulonic acid; D/C Iv fluids

Stat cephalexin, oral, continuous

Influenza vaccine

Pneumococcal vaccine

Consult dietitian

Pancreatic enzymes, oral, continuous

Genetic counseling

*Follow up at 2-3 months

PRIMARY DIAGNOSIS:

Cystic fibrosis

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