

Commentary on Management of Respiratory Problems in Pediatric Cardiac Patients

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Asthma in the Context of Cardiac Disorders

Asthma, as a comorbidity in the presence of cardiac disorder, can be a very annoying issue. In contrast to the patient's cardiac disease, it might not feel like a major problem, especially when considering some cyanotic heart conditions, but in reality, having a daily cough can be very concerning for families, affecting their kid's quality of life, sleep, and daily activity. This requires attention, early evaluation, and intervention by an expert [1].

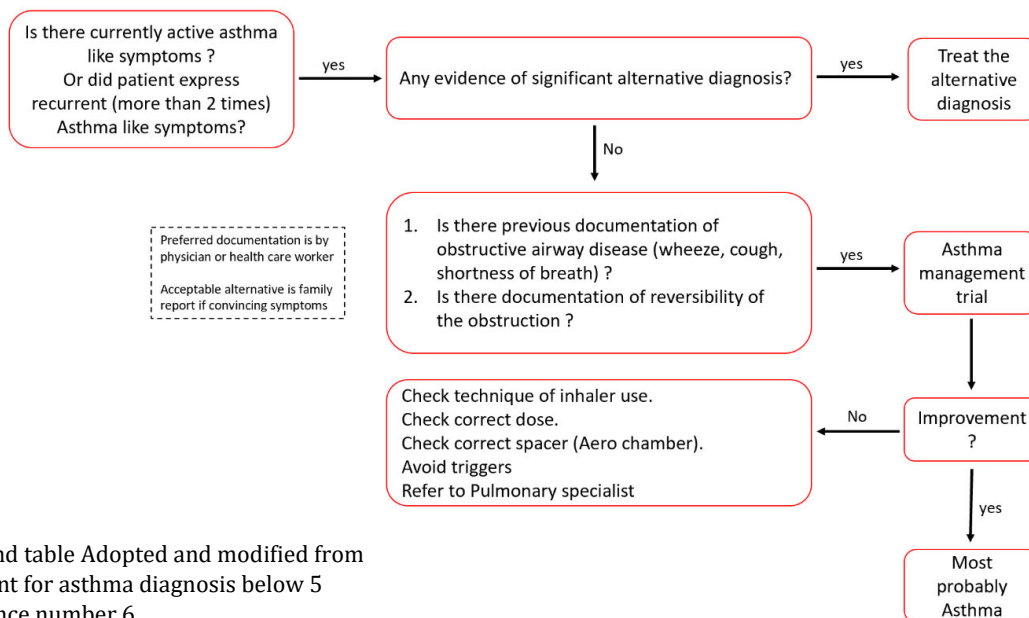
As highlighted before, the word "asthma" equals the reversibility of airway inflammatory disease. With the advances in studying asthma, the terminologies have been changing rapidly; currently, we are specific in linking the phenotype to the endotype as management strategies are changing and advancing, it is critical to understand some basics to achieve the best control of the disease, for example by identifying which cytokine is responsible for the patient's asthma symptoms will tailor his management strategy and plan from simple inhaled corticosteroids inhalers up to biological treatment if required in some cases [1-3].

This will take us to the very basic science behind asthma and even beyond this. As we all know, the most common symptom of childhood asthma is a dry cough followed by a wheeze, and to make it a bit complex, cough receptors are not only in the lower airway! They are present in your ears, upper airway, esophagus, stomach, and the diaphragm; some are even in the brain itself. In other means, asthma phenotype varies, and the final clinical picture that is presented in front of our eyes can not be managed in the same way and needs further and deep assessment to evaluate the mediating cytokine or

immunoglobulin and base the treatment strategy on it [1].

It is challenging to establish asthma diagnoses in children, especially those under five years old, as objective testing to diagnose it is pretty challenging, especially in the context of other significant disorders such as complex heart diseases. Confirmation of asthma requires documentation of reversibility by testing the lung function. While regular Pulmonary Function testing is not feasible for those 5 years of age, accessibility to testing is not easy in other centres. Another challenging factor in cardiac patients is that some older than five will not be able to perform spirometry due to their generalised weakness and heart condition. Asthma is not a cough; a cough can happen because of many conditions, while asthma can present because of different aetiologies. Some have an allergic background, while others are mediated by infections, which make it tricky to control and treat. That's why, in the original article, we focused on trying to give a pathway for differentiating asthma from other asthma-mimicking conditions in the context of complex cardiac conditions utilizing a modified algorithm like the displayed later Flowchart [4,5].

For example, if a 1-year-old patient has a Tetralogy of Fallot had his cardiac defect fixed and expected to do well from a cardiac perspective, surprises you with his recurrent admissions to the hospital because of an increment in his baseline work of breathing and coughing, he should undergo further questioning about the cough and the recurrent respiratory illnesses, for example, if his cough is mainly post feeding, you need to consider alternative diagnosis such as "Reflux, Swallowing incoordination, or even possibility of congenital defects such as laryngeal cleft. This is what has been displayed in the flow chart as an alternative diagnosis.



Asthma alternative diagnosis

signs/symptoms	Possible diagnosis
Recurrent nasal discharge	Allergic rhinitis, Primary ciliary dyskinesia
Stridor, noisy breathing change with position	Upper airway anomalies. Infection laryngomalacia
Acute sudden onset	Foreign body aspiration
First wheeze	Bronchiolitis
Premature birth, needed prolonged oxygen	Bronchopulmonary dysplasia
Chest x-ray abnormality abnormal chest shape	Congenital lung malformation
Chronic productive cough	Bronchiectasis, immunodeficiency, Ciliary dyskinesia, Cystic fibrosis
Cough increase with position change, related to feed	Gastroesophageal reflux disease
Dysphagia, intolerance of solid food	Eosinophilic esohagitis
Chocking with feed, noisy breathing after feed	Swallowing issues, laryngeal cleft

Then if you didn't identify any other treatable condition and remain with asthma, a trial of proper inhaled corticosteroid for a proper period of time, which usually 3 months, should be initiated and assess the response, this is in particular for those who can not perform Pulmonary Functions testing because of age limitation or sometimes weakness and technical limitation [6,7].

One important factor in assessing and managing asthma is avoiding the triggers; this starts by identifying the possible triggers for each patient, which, by the way, differ and are not the same for everyone, so skin prick testing or other modalities of identifying possible exacerbating factors might be required to avoid future exacerbations [7].

Regarding management, it is critical to pay attention to the details of the best treatment choice. In the original article, we spoke about multiple drugs of choice which can be used.

The traditional drug of choice for most caring physicians would be an inhaled corticosteroid through metered dose inhalers, and sadly, on the other hand, it is very common to find that parents don't use the inhalers in the proper technique, comply poorly, and have massive confusion of each drug rule in their kids' condition. For this reason, If the patient's asthma can be controlled with one drug, you need to use only one drug and minimize the burden of families caring for kids with plenty of medications and devices [7,8].

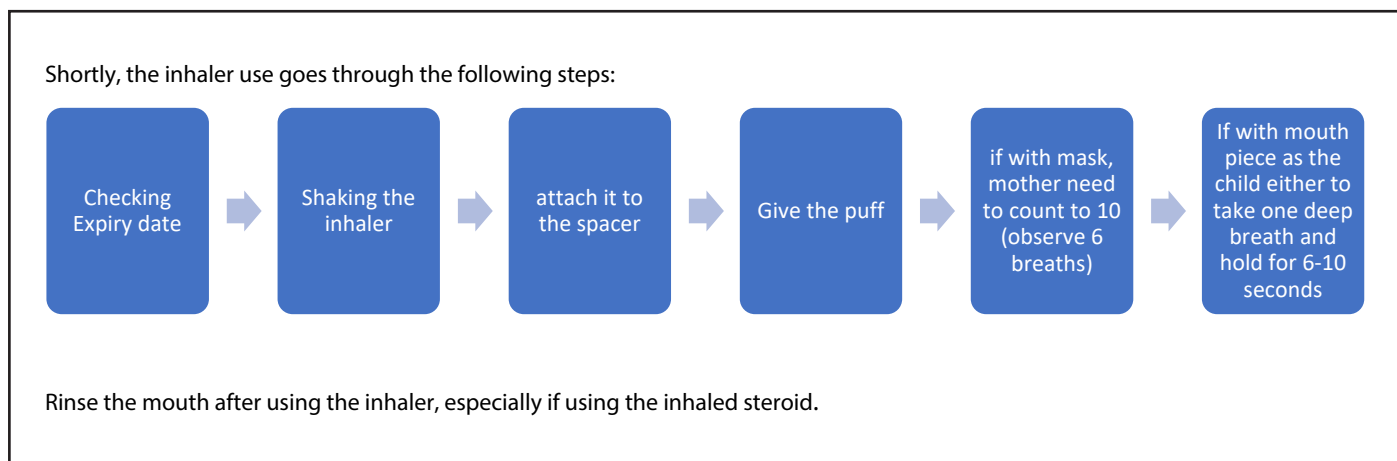
The right technique for using inhalers in the pediatric population includes using the metered dose inhaler and a spacer "Chamber," which is a plastic tube that allows smooth transmission of the medication to the lower airways. The spacer has different sizes and types based on age. For those below 4 years of age, a spacer with a mask (Figure 1) is recommended, while for those above 4 years, it is recommended to use a spacer with a mouthpiece (Figure2) [8].



Figure 1. MDI attached to spacer with mask piece (<4years).



Figure 2. MDI attached to spacer with mouth piece (>4years).



What About Family Education?

Families of patients with underlying asthma condition in addition to their complex cardiac disease, are in need of education and stress on the technique part to ensure delivering the drug to the patient in the best way.

Educating parents start from inpatient setting, if a decision is made to start metered dose inhaler with spacer, then the right spacer should be utilized in inpatient setting and initial doses can be given by health care worker while care givers watches and learn, then before going home the mission can be handled completely by the parents under supervision to

ensure the right technique is being utilized.

Giving the parents visual material and access to online education material will help them memorize whenever they forget the technique and which medication can be used as a rescue and which can be used as maintenance.

A written asthma action plan (**Figure 3**) should be provided, that includes the medications, which should be color coded, when to use the rescue medication, when to increase the dose, and when to call emergency or visit their doctor is another essential step [8,9].

Color	Section	I AM:	MY PEAK FLOW READING:	Medication Instructions
Green	GO: Maintain your medicine.	Breathing easily. Not coughing or wheezing. Working, sleeping and playing easily.	80% to 100% of my personal best.	My doctor recommends these medications: _____ My doctor also recommends that I: _____
Yellow	CAUTION: Step up your medicine.	Coughing, wheezing or feeling shortness of breath. Feeling tightness in my chest. Waking up at night. Experiencing the first signs of a cold.	50% to 79% of my personal best.	My doctor recommends these medications: _____ My doctor also recommends that I: _____
Red	DANGER: Your asthma is getting worse fast.	Not getting relief with my medication. Breathing hard and fast. Struggling to talk.	Less than 50% of my personal best.	My doctor recommends these medications: _____ My doctor also recommends that I: <u>Get medical treatment now.</u>

Figure 3. Written asthma action plan.

Special Cases Related to Complex Cardiac Conditions: Plastic Bronchitis

Plastic bronchitis, which is a very rare fetal condition, can be seen in the context of two conditions: severe uncontrolled allergic asthma and, on the other hand, complex cardiac conditions. Most reports link Fontan surgery to the condition, and to simplify the understanding of the cast formation in the airway in cardiac conditions, it happens more when you change the natural vascular pathway from its original pathway and you change the physiological pathway, that's the time when lymphatic drainage doesn't follow so it finds the nearby airway as a space for clearing and accumulates there [10,11].

As mentioned in the original article, the problem with the conditions is related to the pathology and nature of casts formed, which are acellular in nature and make them less responsive to medical therapy. Despite that, from our experience with a lot of cases, it should be tried as a first line of therapy. This includes Nebulized saline with different concentrations of inhaled steroids and Short-acting beta-agonists, in combination with mucolytic medications and mucus-removing medications such as Acetylcysteine and Pulmozyme. One more drug that can be nebulized in this condition is Tissue plasminogen activator and heparin derivatives, and rising case reports suggest possible responses [12].

In optimum situations, a heart transplant can definitely solve the whole problem, but we all know the limitations in availability, access to service and lack of donations worldwide. While looking at other surgical options, we explored the rule for closing the source of trouble by embolization of the thoracic duct or ligating it, which can work as a bridge toward reaching the ultimate treatment in severe conditions; it doesn't come without costs. and complications such as chronic diarrhoea and abdominal and lower limb swelling [12].

Bronchoscopy, if considered during obstructive events to remove the casts, should be considered with high risks during the operation, keeping in mind that casts can act as foreign bodies and block several levels of the airway, causing more problems when removed than when left a side. When preparing for the procedure, you will need combined rigid and flexible scopes, medications that might help loosen the casts, prepare all forceps and FB retrieving tools that might help the patient during the procedure, and it is recommended if available to utilize cryoprobe as these casts are usually friable [13].

In Summary, although cellular types of casts are reported to be less responsive to nebulized treatments, the author still believe it should be given a chance in the management of it. As some case reports showed some positive responses specially to nebulized Tissue plasminogen activator. In life-threatening situations, such as if a known patient presents

with acute respiratory failure and requires direct intervention, bronchoscopy intervention can be considered, but keeping in mind the difficulty in removing the branching casts and preparing all the required tools with the most expert persons in Rigid and Flexible scopes, with special probes such as cryoprobe if available.

Other surgical interventions, such as thoracic duct ligation embolization, might buy you some time and, as described earlier, don't come without complications; a final surgical correction of the underlying cardiac lesion will be the definitive treatment or heart transplant.

Lastly, plastic bronchitis is a nightmare for physicians taking care of complex cardiac conditions, not only because of treatment difficulties but also because one event can be enough sometimes to cause mortality.

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