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Revisiting Bronchiectasis care in Pakistan- are we following the progress made in developed countries?

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Bronchiectasis affects approximately 500,000 patients in the United States of America. Estimates in Pakistan are difficult due to the lack of registry and recognized diagnostic therapeutic pathways. The cost of bronchiectasis is believed to be more than the combined costs of Chronic Obstructive Pulmonary Disease (COPD) and asthma in developed countries. However, Pakistan still lacks the ability to quantify costs related to bronchiectasis. Attempts have been made to recognize pediatric bronchiectasis research as a priority through the Pakistan Pediatric Association.[1] Additionally, Pakistan is the 5th most populated country in the world, with pneumonia being the leading cause of death. It is quite possible that many patients who die from pneumonia may have underlying bronchiectasis, but it is not yet diagnosed.

Bronchiectasis is believed to be highly prevalent in adult pulmonary clinics in Pakistan. Overall, it is believed to be a result of progression of widely prevalent COPD and asthma in patients. There is a pandemic of infectious diseases, lack of diagnostic capabilities, and as a result, many patients seem to lack specific treatments necessary to treat their underlying condition. There seems to be no current priority for bronchiectasis treatments among the pharmaceutical industry, and there are no research trials to advance bronchiectasis care in Pakistan.

Authors in this paper noted that more than 90% of bronchiectasis diagnosis in Pakistan was from Tuberculosis (TB) compared to the rest of the world, where it is idiopathic. Most patients had a duration of symptoms between 5-10 years. They concluded that Post TB bronchiectasis was the leading cause of non-cystic fibrosis bronchiectasis in Pakistan, and Pseudomonas was the common bacteria prevalent associated with respiratory failure. Spirometry showed obstructive ventilatory impairment and the most significant complication was noted to be hemoptysis.[2]

Another review noted that in Pakistan, TB is endemic and caused 87% of Chronic Pulmonary Aspergillosis (CPA). Furthermore, 12% of bronchiectasis patients had Allergic Bronchopulmonary Aspergillosis (ABPA). 50% of patients had simple aspergilloma, followed by 45% having cavitary aspergillosis.[3]

A recent review from respiratory experts from Agha Khan University (AKU) in Karachi mentions developing guidelines for general practitioners across Pakistan. Authors recognized challenges across timely referrals to pulmonary specialists, as well as adopting recommended practices that can be implemented locally. Various factors have been quoted to have a direct impact on bronchiectasis care such as socio-demographic factors, cultural traditions, and financial avenues.

However, the impact of these practices are still difficult to assess as there are essentially no medications or diagnostic facilities to navigate initial work up or treatment for bronchiectasis patients.[4]

Little is published and known of microbiology practices in bronchiectasis care in Pakistan. This is complicated with the lack of research and infrastructure to timely diagnose, investigate, and appropriately treat according to the guidelines of care. Authors investigated bacterial prevalence and resistance at AKU, Karachi, Pakistan, from 2016-2019. Sputum was collected during acute exacerbation. 345 positive cultures from 160 patients were evaluated. Pseudomonas aeruginosa (PA) was found in the vast majority of these specimens. High prevalence of microbial resistance to commonly used drugs was observed. Authors recommended the need for community level interventions to help recognize infections early to help triage the use of antibiotics.[5]

Authors in this paper highlight surgical resection of localized bronchiectasis. They described localized bronchiectasis in the younger cohort with a mean age of 38 years. Many patients had unilateral involvement and tuberculosis was highly prevalent. They also reported poor lung function of 1.5L, report pneumonectomy in 82 patients, lobectomy in 228 patients, and 88 patients underwent segmentectomy between 2006-2017. The male to female ratio was 2 to 1. These estimates are very different from the demographics of the western world, and the treatment approach is also quite rare.[6] Similar approaches for pediatric lobectomy via thoracotomy have been reported from 2013-2019, which is very uncommon in other parts of world in that age group.[7]

Significant advancements have been made in bronchiectasis diagnosis and management in the last 10 years. Updated guidelines were published in the European Respiratory Society (ERS) and the British Thoracic Society (BTS).[8,9] The American Thoracic Society (ATS) has plans to have an American bronchiectasis guideline. A national bronchiectasis care center network is being developed across America. This will lead to a more robust patient data and registry. Europe has established a network of centers who participate in the EMBARC registry. Many Asian countries are participating with the EMBARC network, such as India and China. This has a global impact in bringing research and therapeutics across the world. Pakistan is not part of this registry. There is heightened interest in new drug development targeting bronchiectasis pathogenesis. Now it is well recognized that bronchiectasis has an inflammatory component, either neutrophilic or eosinophilic. New drug compounds showed that, for the first time in clinical trials, Dipeptidyl Peptidase 1 inhibitors (DPP1) have significant benefits in patients with neutrophilic bronchiectasis. If approved by the Food and Drug Authority (FDA), this will be the first compound given approval in the treatment of bronchiectasis. Emerging concepts showed that inhaled steroids are only beneficial in patients with eosinophilic bronchiectasis. This is clinically important as steroids can worsen the Non-tuberculous Mycobacterium (NTM) disease, which is quite prevalent in bronchiectasis patients. Biologic drugs are being investigated to treat eosinophilic bronchiectasis. Genetic development pathways determine the etiology and can target basic defects. In Cystic Fibrosis (CF), highly effective drug modulators have revolutionized the care of CF patients. For the first time in decades, it has led to more adults living with CF, as a result of improved life expectancy. CF modulator drugs lead to the reduction in frequency of exacerbations and hospitalization, thus preserving the quality of life. CF modulators are not available in Pakistan. Sweat testing has led to an increased diagnosis across adults who have minor versions of CF that can present well beyond 18 years of age. In adult and late diagnosis of CF, patients can present with isolated infertility, pancreatic insufficiency, liver disease as mono-organ disease, and can have preserved lung function. In collaboration with pulmonologists across Pakistan, it has been discussed that CF patients are under-diagnosed. Pakistan does not have sweat testing or genetic testing to recognize and correctly diagnose these late presentations.

Latest developments across the western world, such as spirometry, lung clearance index, nasal nitric oxide, newborn screening, genetic counseling has led to an increase in education, along with screening for genetic diseases. Treatment availability is also extremely limited in Pakistan. There are no guidelines to guide clinicians. A major component of treatment for bronchiectasis is believed to be airway clearance therapy (ACT). Pakistan, however, has no equipment for ACT. There are no physiotherapists who are trained to deliver this therapy. The cost is quoted to be a big factor with this limitation, as common ACT equipment averages several lakh Pakistani rupees. Alternative to these modalities could be breathing techniques, which could potentially bridge this gap; however, there is a lack of education and training available in Pakistan for these cost-effective measures of ACT. There are no standardized aerosols or inhaled antibiotics available to treat bronchiectasis patients. The pharmaceutical industry does not seem to have a priority on manufacturing these therapies. There are no standardized algorithms around antibiotic selection or duration during periods of exacerbation. Hospitals do not have management pathways specifying collection of microbial cultures, or monitoring with spirometry to gauge recovery to baseline.

In conclusion, due to advancements in technology, news and views go around the globe easily. Gone are the days when patients would not have known about these

treatments, or clinicians would lag behind these recent developments. Pakistan produces world-class clinicians who are able to perform at a very high-level when given appropriate resources. Due to patients' access to social media, they are able to seek expert consultations across the world. Lack of treatments and diagnostic equipment could allow them to quickly recognize the limitations they may be facing in Pakistan. Similarly, clinicians also get professional advice from colleagues in western countries, where there are significant advancements as compared to what they may have available. Pakistan should make bronchiectasis diagnosis and treatments a national priority. There seems to be a desperate need for acquisition of diagnostic equipment such as sweat chloride, genetic testing, nasal nitric oxide, lung clearance index, high resolution CT chest (HRCT), microbiological cultures to include microbiome technology, and pulmonary function testing. It is of utmost importance to design a patient registry and become part of EMBARC. This will help bring the latest clinical research trials as well as therapies to Pakistan. Channels are available to mitigate costs and collaborate with appropriate agencies to provide life-saving medications in CF. These steps can save lives and prevent high mortality rates from infectious diseases. Development of protocols around infection control, outpatient and inpatient algorithms, and clinician education can help standardize treatment and improve outcomes. These resources could also assist clinicians to diagnose patients accurately, and provide appropriate treatments necessary for their disease.[10-14]

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