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Review Article

## Strengthening Healthcare Access in Nigerian Tertiary Institutions: A Review of the Tertiary Institution Social Health Insurance Programme

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#### **ABSTRACT**

Health is a critical factor that cannot be overemphasized in the human and social-economic development of any nation. Access to healthcare plays a vital role and is essential for maintaining and improving health. The Tertiary Institution Social Health Insurance Programme (TISHIP) was established by the National Health Insurance Authority (NHIA), and aims to provide affordable and quality health coverage for students in universities, polytechnics, colleges of education, and other post-secondary institutions.

TISHIP is a laudable initiative, designed to reduce out-of-pocket healthcare expenses and protect students from catastrophic medical expenses through pooled contributions. The programme offers a wide range of preventive and curative services without making payments at the point of accessing care. However, various studies have reported mixed findings about the awareness, knowledge, utilization, attitudes and perception about the programme.

Although the NHIA Act of 2022 makes health insurance coverage mandatory for all Nigerians as a means of achieving Universal Health Coverage (UHC), there has been a lack of enforcement, creating major setback to the its implementation. The implementation of TISHIP seems voluntary and at the discretion of each institution, with many having not yet subscribed to it, while some operate it inconsistently.

Hence, the implementation challenges need to be tackled and addressed. Strengthening the programme will require a multi-sectoral approach involving all the stakeholders including NHIA, institutional authorities, HMOs, healthcare facilities and professionals, and student bodies. To improve the programme's effectiveness, efforts should focus on increasing student awareness, strengthening human resources, improving service delivery, and enhancing monitoring and evaluation. It is also essential to ensure transparent fund management, set a premium that is sufficient and economically sustainable, make the programme mandatory, and establish consistent implementation frameworks across all tertiary institutions in the nation.

Keywords: Healthcare, Access, Tertiary Institutions, TISHIP, Nigeria

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#### **INTRODUCTION**

The World Health Organization (WHO) defined health as "a state of complete physical, mental, and social well-being and not merely the absence of disease or infirmity" <sup>1</sup>.

Health is not just a basic necessity; it is essential for individuals' overall well-being, and a crucial means for the pursuit of activities that will enhance human welfare and self-actualization <sup>2,3</sup>. Health is a critical factor that cannot be overemphasized in the human and social-economic development of any nation. Access to healthcare plays a vital role and is essential for maintaining and improving health.

Social health insurance is one of the strategies adopted by the Nigerian government to extend health care delivery to everyone, and the National Health Insurance Authority (NHIA) aims to play a leading role of enhancing access to healthcare services through the development and promotion of various health insurance programmes for different population groups and citizenry across the nation <sup>4</sup>.

Nigeria has a growing population of undergraduates spread across the nation <sup>5-7</sup>, and students in these tertiary institutions occupy a demographic stratum, which cannot be overlooked.

Health and education are part of the basic human needs, and both are fundamentally linked. Education provides opportunities for better health, while poor health can hinder educational attainment <sup>8</sup>. Access to quality healthcare remains a critical factor influencing student well-being and academic performance. Although young persons are generally presumed to be in good health, a number of them face health-related challenges and require health care services, which if not adequately addressed will lead to attendant complications.

## The Tertiary Institution Social Health Insurance Programme

The Tertiary Institution Social Health Insurance Programme (TISHIP) is established under the NHIA to provide social health protection for students in tertiary institutions, which include universities, mono/polytechnics, colleges of education and other post-secondary educational institutions. TISHIP is a social security system whereby the healthcare of students in tertiary education institutions is financed through funds created by pooling the contributions of students <sup>4</sup>. It was part of the policy framework when the NHIS was launched in 2005. Its primary aim is to meet the healthcare needs of students in tertiary schools, who make up a considerable portion of the national population, particularly students who have

reached the age of 18 years, and are not eligible for coverage under the public sector scheme as dependents of enrolled parents <sup>4</sup>, while not forgetting those whose parents are not under the public sector scheme, hence, cannot be enrolled as dependents.

TISHIP aims to provide quality and affordable healthcare coverage to all students in Nigerian tertiary institutions while taking into account the peculiarities of their health needs. The programme seeks to protect students from the financial hardships of huge medical bills, ensure the availability of funds to the tertiary institution health centres for improved services, reduce out-of-pocket payments and protect students from catastrophic health expenditures 4. Although the National Health Insurance Authority (NHIA) Act of 2022 makes health insurance coverage mandatory for all Nigerians as a means of achieving Universal Health Coverage (UHC) 9, there has been a lack of enforcement 10, creating major setback to the Act's effective implementation. The implementation of TISHIP seems voluntary and at the discretion of each institution, with many having not yet subscribed to it, while some operate it inconsistently.

#### Membership, Eligibility and Funding

Membership is for full and part-time students of Federal, State and Private Tertiary education Institutions who are not on any form of mandatory health insurance <sup>4</sup>.

It is a contributory scheme; hence students need to pay for TISHIP upon resumption of each academic session as part registration process. Funds from charitable organizations, corporate initiatives, government interventions, and donations can also provide subsidies to support enrolment costs where available <sup>4</sup>.

The lowest premium set by the NHIA is Two Thousand naira (N2,000:00k) per session <sup>11</sup>, though many institutions adjust their own premium up due to the prevailing economic realities.

#### The Benefit package and Scope of Coverage

TISHIP provides services that are within NHIS scope of coverage for all contributory social health insurance schemes and programmes <sup>4</sup>, which consists of promotive, preventive, curative and rehabilitative services at both primary and secondary levels of care, provided by public and private health facilities accredited by the NHIA.

Under this programme, beneficiaries are exempted from making co-payments at the point of accessing healthcare services unlike that of the public sector programmes. Additionally, students can access care whether the school is in session or on break and in emergencies, providing continuous coverage and uninterrupted care throughout the year in order to address health needs that may arise during holidays or outside the regular academic calendar.

#### Stakeholders of the Programme

The key stakeholders involved in the programme include NHIA, tertiary institutions, Health Maintenance Organizations (HMOs), Student Union, State Social Health Insurance Agencies (SSHIAs), healthcare facilities, and regulatory bodies of tertiary institutions <sup>11,12</sup>. The Programme is managed within the institution by the TISHIP committee comprising of representatives of all the stakeholders.

#### **Impact**

TISHIP is a laudable initiative aimed at providing quality and affordable healthcare coverage to students in Nigerian tertiary institutions. However, various studies have reported mixed findings about the awareness, knowledge, utilization, attitudes and perception about the programme <sup>13-18</sup> Anecdotal reports from Health Maintenance Organizations (HMOs) also suggest considerable levels of utilization by students, highlighting the need for datadriven evaluation of the programme's actual effectiveness and impact.

#### Challenges

The implementation of GIFSHIP is faced by several challenges. These include inconsistent implementation across institutions, with many operating the programme in different ways, while some are yet to subscribe to it at all. Poor sensitization and enlightenment of students about the programme, leading to low awareness and participation. Inefficient service delivery, inadequate infrastructure, poor staff attitudes and long waiting times, which further discourage utilization. Additional barriers include insufficient funding, lack of transparency in the collection and remittance of funds, and poor TISHIP management <sup>18-</sup>

#### **CONCLUSION**

In conclusion, TISHIP plays a vital role in improving healthcare access for students in Nigerian tertiary institutions. However, the implementation challenges need to be tackled and addressed. Strengthening the programme will require a multi-sectoral approach involving all the stakeholders including NHIA, institutional authorities, HMOs, healthcare facilities and professionals, and student bodies. To improve the programme's effectiveness, efforts should focus on increasing student awareness, strengthening human resources, improving service delivery, and enhancing monitoring and evaluation. It is also essential to ensure transparent fund management, set a premium that is sufficient and economically sustainable, make the programme mandatory, and establish consistent implementation frameworks across all tertiary institutions in the nation.

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Review Article

### Parkinson's Disease and Its Impact on Quality of Life

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#### **ABSTRACT**

**Background**: Parkinson's disease (PD) is a progressive neurodegenerative disorder which significantly impacts on the quality of life (QoL) of affected individuals. All the components of QoL (physical functioning, psychosocial, cognitive, and environmental) can be affected by the symptoms of Parkinson's disease, side effects of treatment, and sociocultural factors.

**Methodology**: This study is a literature review of the quality of life in patients with Parkinson's disease. Excerpta Medica database (EMBASE), Medical Literature Analysis and Retrieval System Online (MEDLINE), Public Medline (PUBMED), Health Inter-Network Access to Research Initiative (HINARI) and Google Scholar databases were searched and scholarly articles on Parkinson's disease and QoL were reviewed for the purpose of the study.

**Results**: Parkinson's disease is primarily caused by the loss of dopaminergic neurons in the substantia nigra which result in motor and non-motor symptoms (such as cognitive function) with profound negative impact on the QoL. Also, with progression of the disease, and worsening of symptoms, an attendant decline has been observed in the QoL of patients with Parkinson's disease. Family and social support is crucial in the management of the condition.

**Conclusion**: PD significantly impacts on the QoL of patients and should be assessed for. Recognition of challenges in the lives of patients with PD is important in their management in addition to drug therapy. Family and social support including occupational and financial support are relevant in management.

**Keywords:** Neurodegenerative disorder, Parkinson's Disease, Quality of Life, Cognition, Physical Functioning.

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#### **INTRODUCTION**

Parkinson's disease (PD) is a progressive neurodegenerative disorder with significant impacts on the quality of life (QoL) of affected individuals. All the components of QoL (physical, psychological, social, and environmental) can be affected by the symptoms of Parkinson's disease, side effects of treatment, and sociocultural factors. Parkinson's disease is primarily caused by the loss of dopaminergic neurons in the substantia nigra which result in motor and non-motor symptoms with profound negative impact on the QoL.<sup>1-4</sup> PD has a prevalence of 1-2 per 1,000 with increment in age above 60 years reaching about 4% in the elderly population.<sup>2-4</sup> PD occurs in the young, though rare, and may be familial Affecting multiple family members in which case it's usually associated with genetic predisposition.<sup>5-7</sup> The condition is characterized by motor dysfunctions such as resting

tremors, rigidity, bradykinesia, gait freezing and postural reflex abnormalities as well as non-motor dysfunction such as cognitive decline, depression, anxiety, sleep disturbance and fatigue which impact negatively on the QoL.<sup>1,2,3,8</sup>. The objective of the study was to review the range of factors that negatively impact the quality of life of patients with Parkinson's disease. A narrative literature review was adopted for the purpose of this study.

#### **DISCUSSION**

For research purposes there are validated tools for use in objectively measuring the QoL in Parkinson's disease. These include the Parkinson's Disease Questionnaire (PDQ-39)<sup>10</sup> and/ or its shorter version (PDQ-8),<sup>11</sup> which assess domains such as mobility, activities of daily living (ADL), emotional well-being, stigma, social support, cognition, communication, and bodily discomfort.<sup>10-12</sup>

#### Physical and functional impacts on quality of life

Parkinson's Disease (PD) significantly impacts an individual's quality of life through a range of physical and functional challenges. The degeneration of dopaminergic neurons in the substantia nigra, a hallmark of PD, leads to a variety of motor symptoms that directly impair daily activities and overall well-being<sup>13</sup>. These physical manifestations include bradykinesia (slowness of movement), rigidity (stiffness), tremor, and postural instability, which collectively hinder mobility, balance, and coordination<sup>13</sup>. Consequently, individuals with PD often experience difficulties with fundamental tasks such as walking, dressing, eating, and writing, leading to a loss of independence and increased reliance on caregivers. Beyond motor symptoms, PD also presents significant non-motor impacts, which can be equally or even more debilitating. These include fatigue, pain, sleep disturbances, autonomic dysfunction (e.g., constipation, orthostatic hypotension), and neuropsychiatric symptoms such as depression, anxiety, and cognitive impairment.<sup>14</sup> The combination of these physical and functional limitations, coupled with the progressive nature of the disease, can severely diminish an individual's ability to participate in social activities, maintain employment, and engage in hobbies, ultimately leading to a reduced quality of life and increased social isolation.<sup>14</sup>

#### Cognitive impairment

About 20-50% of patients with Parkinson's develop Mild Cognitive Impairment (MCI), progressing to dementia in 30-80% of cases over time, leading to a sharp decline in judgment, memory, and executive function hence reducing QoL.15,19 Studies have shown that cognitive dysfunction is a common non-motor symptom in individuals with PD, affecting a substantial number of patients, including Nigerians with the disease. 15,16,18 These cognitive changes can manifest as difficulties in attention, executive function (such as planning and problem-solving), processing speed, and visuospatial abilities. 15,19 For instance, the Community Screening Instrument for Dementia (CSID) has been utilized to assess the frequency, pattern, and predictors of cognitive impairments in PD patients, highlighting the diverse nature of these deficits.<sup>15</sup> Furthermore, tools like the Six-item Cognitive Impairment Test (6CIT) have been validated as screening tools for cognitive dysfunction, emphasizing the need for effective assessment in this population.<sup>18</sup> The progression of cognitive decline in PD can have a profound effect on the quality of life for patients and their caregivers, often imposing a greater burden than the motor symptoms of the disease.<sup>19</sup> This underscores the importance of understanding the multifaceted impact of Parkinson's disease on cognitive function for comprehensive patient care.

#### Sleep disturbances

Sleep disturbances are a pervasive and significant nonmotor symptom in Parkinson's disease (PD), often having a considerable negative impact on patients' quality of life. 20 These disturbances are complex, arising from a combination of factors including the neuropathophysiological of PD itself, primary sleep disorders, co-occurring medical or psychiatric conditions, and the effects of anti-Parkinson's medications.<sup>20</sup> Common manifestations of sleep dysfunction in PD patients include insomnia, characterized by difficulty initiating or maintaining sleep, and excessive daytime sleepiness.20 Other sleep issues frequently observed are sleep fragmentation, restless legs syndrome, and REM sleep behaviour disorder, where individuals physically act out their dreams.<sup>20</sup> The evaluation of these disturbances is crucial for effective management, though treatment strategies are still an area of ongoing research, often drawing on clinical experience and studies from other geriatric populations.<sup>20</sup> Addressing sleep problems in PD is vital for improving overall well-being and mitigating the substantial burden they place on both patients and their caregivers.

#### Autonomic dysfunction

With involvement of the Autonomic nervous system, patient may develop symptoms like constipation, urinary incontinence, sialorrhea, orthostatic hypotension, and sexual dysfunction. The article "Autonomic Dysfunction" by Sánchez-Manso *et al.* makes a brief mention stating that "Other common manifestations are related to postural tachycardia syndrome (POTS) or changes seen with Parkinson disease and other parkinsonisms".<sup>21</sup> This indicates that autonomic dysfunction, particularly issues like POTS or other general autonomic changes, are observed in individuals with Parkinson's disease, all of which can impact negatively on the QoL.<sup>21</sup>

#### Depression and anxiety

The sad reality of living with a chronic progressive condition and the hormonal imbalance resulting from Parkinson's disease significantly increase the risk of Mental health disorders like depression and anxiety, not only does it affect the motor system, but significantly because of the high prevalence of non-motor symptoms such as depression and anxiety. As highlighted by Al-Khammash et al., these neuropsychiatric issues are extremely common in individuals with PD and can severely diminish their overall well-being.1 A systematic review and meta-analysis by Zhao et al. further reinforces that depression and anxiety are prevalent and substantially contribute to a poorer quality of life in PD patients.2 Garcia-Ruiz et al. emphasize that these mood disorders are not merely secondary reactions to the chronic illness but are integral aspects of the disease pathophysiology itself.8 Therefore, effective management of depression and anxiety is a critical, yet often overlooked, component of holistic care for individuals living with Parkinson's disease, recognizing their significant impact on patient well-being beyond motor manifestations. 1,2,3,8

#### Social and relational impacts

Overtime, patients with Parkinson's disease become dependent on relatives and caregivers both for their daily activities and financial obligations, often extending beyond the direct patient to their family and caregivers. As highlighted by Ogbimi et al. in their case report on Familial Early-Onset Parkinsonism, the socioeconomic impact of the disease is substantial, affecting the patient's independence and placing a considerable burden on caregivers<sup>5</sup>. The need for continuous care and the increasing disability associated with PD necessitate a significant adjustment in close relationships, leading to changes in family dynamics and roles. While Mehanna et al. focus on age cutoffs for Early Onset Parkinson's Disease,6 implicitly, the earlier onset means a longer duration of impact on social and relational aspects, affecting careers, family planning, and peer relationships over a more extended period. The challenges posed by PD, including its motor and non-motor symptoms, can lead to social withdrawal, reduced participation in activities, and strains on interpersonal connections, underscoring the comprehensive social and relational support required for patients and their support networks.

#### Social isolation

Parkinson's disease (PD) has a profound impact on social functioning, frequently leading to social isolation and withdrawal, which can significantly diminish a patient's quality of life.<sup>22</sup> This social withdrawal can be both voluntary and involuntary, often stemming from the interplay of various physical, cognitive, and psychiatric symptoms associated with PD, as well as the perceived stigma of the disease.<sup>22</sup> Motor symptoms like tremors, gait disturbances, and speech difficulties can make social interactions challenging and embarrassing, leading individuals to avoid social situations. Furthermore, non-motor symptoms such as depression, anxiety, and fatigue contribute significantly to reduced social engagement.<sup>22</sup> While Ogbimi et al. discuss familial early-onset parkinsonism and Mehanna et al. focus on age cutoffs for early-onset PD, Ahn et al. specifically highlight that individuals with PD may reduce their social activities due to these complex and debilitating effects, emphasizing the critical need to address social isolation in comprehensive PD care.5,6,22

#### Communication difficulties

As Parkinson's disease progresses, patient may develop hypophonia or slurring of speech which can hinder communication and contribute to loneliness and social isolation. As discussed by Prenger *et al.*, individuals with PD

often experience disruptions in their ability to produce emotional facial expressions (known as facial masking) and emotional speech (dysarthria). He beyond production, they also struggle with recognizing the verbal and nonverbal emotional cues of others. These impairments can lead to severe negative social consequences, including feelings of stigma, dehumanization, and loneliness. The communication challenges extend to difficulties in modulating facial expressions based on social context, impacting the ability to maintain positive social relationships. Ultimately, these "social symptoms" of PD, encompassing various communicative changes, can cause major disruptions to social functioning and significantly reduce a patient's quality of life, sometimes even more so than the more recognized motor or cognitive symptoms. He

#### Financial strain

The treatment of Parkinson's disease can be burdensome as it places significant financial costs on patients and relatives due to medications, physiotherapy and occupational therapy, and other indirect cost of the disease, primarily due to the chronic and progressive nature of the condition. Ogbimi et al., in their case report on familial early-onset parkinsonism, touch upon the "socioeconomic impact" of the disease, which inherently includes financial considerations, early onset can lead to a longer duration of disease, potentially increasing lifetime care costs and impacting earning potential.<sup>5</sup> Furthermore, while Ahn et al. primarily review social withdrawal in PD, they highlight that social withdrawal can be associated with various factors including physical, cognitive, and psychiatric symptoms.<sup>22</sup> These symptoms, in turn, can affect a patient's ability to maintain employment or engage in activities that contribute to household income, thereby indirectly leading to financial strain. The comprehensive burden of PD, encompassing both direct medical expenses and indirect costs such as lost wages and caregiver burden, inevitably results in considerable financial hardship for those affected.<sup>5,22</sup>

#### Treatment-related impacts on quality of life

While treatments can improve symptoms, the side effects of some of the medications can be challenging and can potentially reduce QoL. Levodopa is the gold standard for symptomatic treatment of PD, and it is highly effective in improving motor symptoms. This improvement in motor

function can lead to a better quality of life in the short term. However, the long-term use of levodopa is associated with several complications that can adversely affect quality of life. These include the development of motor fluctuations, such as "wearing off" effects where the medication's benefits diminish before the next dose, and dyskinesias, which are involuntary, jerky movements.<sup>24</sup> These side effects can be quite disabling and significantly impact a patient's daily life and overall well-being. Additionally, common adverse effects of levodopa include nausea, dizziness, headache, and somnolence. In older patients, more severe central nervous system effects such as confusion, hallucinations, delusions, psychosis, and agitation can occur.24 Abrupt withdrawal or dose reduction of levodopa also carries a risk of serious complications like parkinsonism hyperpyrexia syndrome.24

#### **CONCLUSION**

PD is a chronic neurodegenerative disease with significant impacts on the QoL of patients. The negative impact is multi-systemic as well as physical, and psychosocial impairment. While the physical symptoms encompass mostly the motor manifestations, studies have established that cognitive impairment is the most common non-motor manifestation. Recognition of challenges in the lives of patients with PD which impact on their QoL is important in their management in addition to drug therapy. Family and social support including occupational and financial support is relevant in management.

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Case Report

## Giant Gall Bladder Stone: A Case Report and Review of Literature

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#### **ABSTRACT**

Gallbladder stones usually exhibit significant variation in size but rarely up to 5cm. Giant gallbladder stones, defined as stones greater than 5cm in diameter or more than 70 grams in weight, are a rare phenomenon and pose a management challenge. Laparoscopic cholecystectomy while being the gold standard of surgically removing gallbladders has a high rate of conversion to open even in expert hands due to the challenges of handling the giant gallstones. These challenges include difficulty grasping the gallbladder with the laparoscopic instruments, difficulty in exposing the anatomy of Calot's triangle due to the size of the stone and extrinsic compression on the Calot's triangle and difficulty in retrieving the specimen. We present the management of our first case of cholecystectomy for a giant gallbladder stone and discuss the choice of surgery.

Keywords: Giant, gallbladder stone, laparoscopic cholecystectomy, Calot, fundus first.

#### **INTRODUCTION**

Gallstone diseases were predominantly seen in the developed world. However, over the last few decades, the incidence in developing countries has been steadily increasing with recent reports placing the incidence of gallstone diseases at 2% to 6% in most African nations. The exact aetiology of gallstones is not known but a multitude of risk factors has been implicated. The most consistent risk factor reported is the female sex. Other risk factors include obesity, rapid weight loss, Caucasian race, age more than 40 years, dyslipidemia, sedentary lifestyle and diabetes mellitus. 4,5

The pathogenesis of gallstones has been studied extensively and it is believed that biliary supersaturation from hypersecretion of cholesterol forms an integral part of the formation of cholesterol gallstones. Other alterations in the hepatobiliary system observed in patients with gallstone include accelerated nucleation, gallbladder dysmotility, and mucin gel accumulation.<sup>6–8</sup>

Gallstones usually exhibit significant variation in size, they may range from 3mm to more than 5cm. Typically, less than 5mm gallstones may not require treatment because they may pass spontaneously through the bile duct without any symptoms. However any gallstone of more than 10mm may obstruct the bile ducts causing jaundice and infection which will necessitate cholecystectomy and common bile duct exploration.

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The size of the stone may also increase the risk of gallbladder cancer. Studies have shown that gallstones greater than 3 cm carry a higher risk for gallbladder cancer.10 Giant gallbladder stone is defined as a gallstone that is more than 5 cm in its widest diameter or weighing above 70g.11-13 The treatment of giant gallstones is challenging because of the difficulty associated with laparoscopic delivery of the gallbladder. Some surgeons believe that this is an indication of open cholecystectomy but this has been disputed with multiple reports of successful laparoscopic cholecystectomy in giant gallstones albeit with some difficulties.11–13 In a developing country like ours where laparoscopic surgery facilities are inadequate and expertise is slowly gaining momentum, removing giant gallstones with open surgery is an easier decision to make. We present a report on our first case of giant gallstone treated with open cholecystectomy.

#### Case report

A 65-year-old woman presented to us with severe right hypochondrial pain of 2 weeks duration which started gradually as a dull ache. It had become severe and colicky and radiated to the right shoulder. There was associated nausea, anorexia and 4 episodes of vomiting. She had a low-grade fever for a few days but it subsided after a course of antibiotics. There was no jaundice.

She had a long-standing history of dyspepsia/ epigastric discomfort of more than 10 years. It was usually precipitated by fatty meals. She had used antacids with minimal relief.

She was neither a known hypertensive nor a known diabetic patient.

On examination, she was an elderly woman, not ill, not in any distress, afebrile, not pale, and anicteric.

The abdomen was flat and moved with respiration. There was a palpable roundish mass at the right hypochondrium that was tender, Murphy's sign was positive. A diagnosis of resolving acute calculous cholecystitis was made.

An abdominal ultrasound scan revealed a huge, rounded gallbladder stone which measured 6cm by 5.5cm across. She was prepared for cholecystectomy. Under general anaesthesia, Kocher's incision was made (figure 1) to deliver the gallbladder which was then removed through the 'Fundus first' approach.

Cut open, the excised gallbladder contained about 100mls of mucinous fluid and a giant dark-coloured stone that measured 6.3cm by 5.5cm and weighed 63 grams. (figure 2).

Her recovery was uneventful. Histology of the excised gallbladder showed features of chronic inflammation and no evidence of malignancy.

Qualitative Chemical analysis of the stone revealed a mixed stone containing calcium, xanthine, phosphate, cholesterol, and bilirubin as the main contents. It was negative for oxalate and urate.



Figure 1: Intraoperative picture of the huge gallbladder, approached through a Kocher's incision



Figure 2: Huge dark-colored stone, the width of 3 fingers, removed from the gallbladder. Measured 6.3 by 5.5cm.

#### **DISCUSSION**

Gallstone diseases constitute a significant burden to the population of North America and Western Europe. It is believed to affect up to 15 % of the population in these parts of the world. In most parts of Africa, the burden of gallstone disease is minimal but it has been increasing in the past few decades.<sup>1–3</sup> The exact aetiology of gallstones is not known but a multitude of risk factors has been implicated. These risk factors include female sex, obesity,

rapid weight loss, Caucasian race, age more than 40 years, sickle cell disease, dyslipidemia, sedentary lifestyle and diabetes mellitus.<sup>4,5,7</sup>

Most gallstones are not symptomatic. Only about 20% of gallstones become symptomatic. It may manifest as symptomatic uncomplicated gallstones when it presents as biliary colic or flatulent dyspepsia. Or it can present as symptomatic complicated and this spectrum includes acute cholecystitis, common bile duct stones, cholangitis, and pancreatitis. Cholecystectomy is recommended for all complicated gallstones. In the presence of symptomatic uncomplicated gallstones, current clinical practice guidelines have no consensus but most surgeons opt for cholecystectomy. 14,15

Giant Gallbladder stone is defined as a gallstone that is more than 5 cm in its widest diameter or weighing above 70g.11-13 Though reports showed that giant gallstones are rare, most of these stones are symptomatic at presentation and require operative treatment. Laparoscopic cholecystectomy is the gold standard for the treatment of gallstone disease but in giant gallstones, there are reports of difficulty in laparoscopic cholecystectomy. This difficulty is attributed to more severe inflammation and thickening of the gallbladder wall, difficulty grasping the gallbladder with the laparoscopic instruments and difficulty in exposing the anatomy of Calot's triangle due to the size of the stone and extrinsic compression on the Calot's triangle.11-13 These anticipated problems have resulted in some surgeons proposing giant gallstones as one of the indications for open cholecystectomy in the current settings and that is why in this index patient, we chose open cholecystectomy. 16,17 However, with improvement in skills and dexterity in laparoscopic surgical practice, there are reports of successful laparoscopic cholecystectomy in giant gallstone disease. It has been shown that laparoscopic fundus first cholecystectomy is associated with fewer complications compared to conventional Calot first cholecystectomy especially in difficult cholecystetomies.<sup>18</sup> So for patients with giant gallstones, the use of fundus first laparoscopic cholecystectomy may be associated with reduced conversion to open cholecystectomy. 18-20 Singh et al 12 reported in year 2020, the removal of a 12.8 x 7cm gallstone as the largest gallstone removed laparoscopically in the world. Xu et al<sup>13</sup> reported emergency laparoscopic cholecystectomy with the removal of

a 9.5cm stone. In Nigeria, Igwe et al reported a series of two patients that had successful laparoscopic cholecystectomy with the removal of an 8 cm stone and an 8.2cm stone.<sup>11</sup>

#### **CONCLUSION**

Giant gallstone diseases are usually symptomatic and may require open cholecystectomy. With the increase in skills and dexterity, laparoscopic cholecystectomy can be performed successfully in patients with giant gallstones. In any case, the 'Fundus first approach' is most suitable.

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Case Report

## Pulmonary Arterial Hypertension in a 14-Year-Old Girl: Diagnostic Challenges and Management

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#### **ABSTRACT**

Pulmonary hypertension carries a high morbidity and mortality especially when presentation occurs late. Due to its subtle manner of presentation and high cost of care, diagnosis is often delayed and treatment is beyond the reach of most affected Nigerians. A 15-year-old girl who presented with easy fatigability of 2 years duration and dypnoea on exertion of 3-weeks duration. She had features of systemic and pulmonary congestion on examination. An echocardiogram done revealed supra-systemic pulmonary pressure with a massively dilated and poorly functional right ventricle. A large secundum ASD with bidirectional flow of blood was also noted. She was commenced on anti-failure regimen and treatment for pulmonary hypertension. Due to high cost of care, combination therapy for pulmonary hypertension could not be sustained and patient eventually succumbed to the illness.

#### INTRODUCTION

Pulmonary hypertension (PHT) is defined as mean pulmonary arterial pressure greater than 25mmHg at rest.¹ The condition is characterized by exercise intolerance in the early stage and right ventricular failure in the late stage.² It is one of the most challenging aspects of cardiology with limited success in management especially with late presentation. Pulmonary hypertension carries a high morbidity and mortality especially when presentation occurs late.³ Its management is very complex, expensive and requires specialized cardiac centres. The diagnosis is often made late due to its non-specific presentation. The index case is a 15-year-old girl that presented with right ventricular failure due to pulmonary hypertension with poor response to medical therapy.

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#### Case Presentation

A 15-year-old girl with easy fatigability of 2 years duration, fast breathing, orthopnoea and paroxysmal nocturnal dyspnoea of 3 weeks duration. Past medical history revealed that from her early childhood she had several admissions for cough and difficulty in breathing. She was observed to be sluggish and not quite as active as other children. However, conditions worsened in the last 2 years when minor daily activities like walking and singing resulted to breathlessness with worsening orthopnoea that requires her leaning forward on a chair to sleep.

#### Clinical findings

She was wasted and cyanosed with grade 4 digital clubbing and generalized pitting oedema.

The PR was 120bpm, BP-90/60mmHg, JVP was 12cm, and apex beat was displaced to 7th intercostal apace along the anterior axillary line. Additional findings included right parasternal heave, S3 gallop rhythm and a pan-systolic murmur loudest at the left lower sternal border. The RR-40cpm, dyspnoeic at rest with bi-basal crepitation. She had ascites demonstrable by fluid thrill with tender hepatomegaly. Her SPO<sub>2</sub>- 70% and on oxygen appreciated to about 80%.

#### Diagnostic assessment

Chest radiograph showed cardiomegaly and on echocardiogram revealed severely dilated right ventricle, flattening of the interventricular septum, a large pulmonary artery, dilated IVC, measuring 20.0mm with poor collapse during inspiration, a large Secundum ASD measuring 39.6mm with bidirectional flow. The mean pulmonary pressure estimated from the pulmonary regurgitant jet was 46.85mmHg and the sPAP estimated from tricuspid regurgitant jet and RAP was 102mmHg (sPAP + RAP). Right ventricular function was poor with TAPSE of 11.0mm and fractional area change (FAC) of 10.0%. The left ventricle appears squashed by the supra-systemic pulmonary pressure; however, the ejection fraction was adequate and no significant left valvular heart disease. Screening for HIV and schistosomiasis were negative. Chest CT scan and Right Heart Catheterization were not done due to financial constraints.

The diagnosis was right ventricular failure 2º pulmonary arterial hypertension with background large ASD (?Eisenmenger Syndrome). Figure 1-4 show the findings on echocardiogram.

#### Therapeutic intervention

Initial treatment was with antibiotics for pneumonia, diuretics and ACE inhibitors for heart failure and Sildenafil for PHT. The D dimer was elevated and as a result patient was commenced on anticoagulation therapy with heparin and warfarin. The SPO<sub>2</sub> improved to 91%, however, pulmonary pressures remained supra-systemic. Patient was discharged at WHO functional class III for follow up in the cardiology clinic. After 6 months of monotherapy with sildenafil, borsetan was introduced. However, due to the high cost of the medications, it was stopped after 2 weeks. Parents who were subsistence farmers were

counselled on the poor prognosis of the child's condition and the need for heart-lung transplants. The condition was also discussed in a webinar session with a US based NGO that partners with some health facilities in Nigeria to provide surgical treatment for children with heart conditions. Due to the difficulty with donors, and the challenges of management, the case did not get the needed attention during the discussions. The patient continued to decompensate with features of systemic and pulmonary congestion with poor response to anti-failure therapy. Patient succumbed to the illness after about a year of treatment.

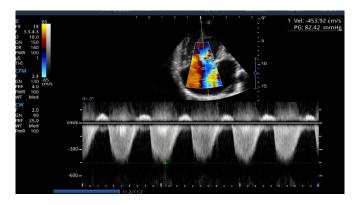


Figure 1: TR jet from Apical 4 Chamber View

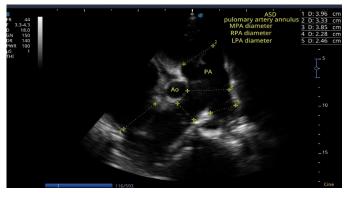


Figure 2: Pulmonary Artery from Parasternal Short Axis View



Figure 3: Right Ventricle versus Left Ventricle

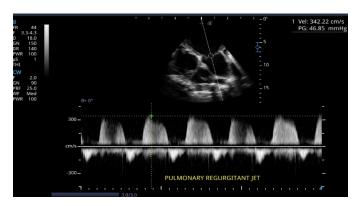


Figure 4: Pulmonary Regurgitant Jet for Mean Pulmonary Artery Pressure Estimation

#### **DISCUSSION**

Pulmonary hypertension (PHT) is a bag of several diseases. It encompasses various conditions that share the common outcome of high blood pressure in the pulmonary arteries, ultimately leading to RV failure and death.<sup>4</sup> The WHO classifies PHT into 5 categories and groups share histology, pathophysiology and likely specific treatment or prognosis.<sup>4</sup> Early stage of the disease often goes unnoticed until irreversible damage has been done to the pulmonary vasculature and the right ventricle. The index case presented in the terminal stage of the disease, a stage that poorly responds to medical therapy.

Pulmonary arterial hypertension (PAH) is defined as a mean pulmonary arterial pressure greater than 25 mmHg at rest, with a normal pulmonary artery wedge pressure less than 15 mmHg and an increased pulmonary vascular resistance greater than 3 Wood/m<sup>2</sup>. There are few studies that have reported the prevalence/incidence of PHT in children and they are mostly in the developed world. The incidence of idiopathic PAH in the national registry from the United Kingdom was 0.48 cases per million children per year and the prevalence was 2.1 cases per million children.<sup>5</sup> In the Netherlands, annual incidence and point prevalence averaged 0.7 and 2.2 cases per million children respectively.6 The prevalence is higher in conditions such as HIV infection, sickle cell anaemia and in congenital heart diseases (CHD).7 The prevalence of PHT in HIV is reported to be about 2,500 times higher than the general population.8

The index case screening for HIV and schistosomiasis was negative. However, a large secundum ASD was discovered on echocardiogram. Atrial septal defects, due to their insidious presentation, can go undetected because these patients do not present with overt heart failure in child-hood due to the fact that the atria are low pressure chambers. Though pulmonary hypertension can occur at any age in life following a CHD, they commonly occur in the 4th decade of life. The condition is also described as Eisenmenger syndrome. It is not quite clear if the pulmonary hypertension was due to the background ASD or if it was idiopathic

A similar case has been reported in the literature in 35year-old women who was known to have OS-ASD measuring 23mm. Patient developed PAH with PASP-85mmHg, MAP-55mmHg and a PVR of 3.38woods. She had combination therapy with ambrisentan and tedalafil for 3 months with significant improvement in her WHO functional class (from IV to II) and her 6-minute walk (110-255m). The response to the combination therapy of endothelin receptor antagonist and 51- phosphodiesterase inhibitor occurred after 3-months.<sup>10</sup> Another case reported was a 19-year-old boy with OS-ASD that was complicated by severe PAH, reported in Nigeria.<sup>11</sup> This case was lost to follow-up after discharge, most likely due to financial constraint. The index case could not afford combination therapy and did not respond to monotherapy. Guidelines for managing patients with PAH that failed to respond to monotherapy state that two or three agent combination therapy with 5-phosphodiesterase inhibitor, endothelin receptor antagonist and prostacyclin analogues should be used.

Several investigations are required to make the diagnosis of PAH due to the fact that there are many other secondary causes. As part of evaluation, patients are required to undergo a right heart catheterization and a vasoreactivity test using inhaled nitric oxide or intravenous epoprostenol during the procedure. Those who respond are commenced on calcium channel blocker, however, our patient did not have a RHC due to lack of funds. There are also limited centres that provide this level of care in Nigeria with few specialists. Additionally, invasive procedures such as RHC require general anaesthesia, and these can be catastrophic for patients with PHT. Mortality can be as high as 23% during some major procedures. 12 A collaborative team has to pre-evaluate, risk assess & optimize and give parents enough time to weigh the risk of procedures. PAH in children without intervention has a

median survival of about 10 months after diagnosis.<sup>13</sup> Heart lung transplantation which is the definitive management in terminal stage of the disease is challenged by availability of donors, lack of expertise, the risk of surgery and cost of care.

#### **CONCLUSION**

Management of PAH requires a team of dedicated experts in the field of cardiology in a specialized centre. The diagnosis is often made late but exertional dyspnoea with no apparent cause can serve as a clue. ECG which is relatively available in most settings and affordable can serve as a ready screening tool

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