

# ME-JAA

Middle East Journal of Age and Ageing

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Volume 8, Issue 2

March 2011

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# Editorial

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*Dr Abdul Abyad*

Chief Editor

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With this second issue of the journal we would like to greet all the physicians and health personnel helping people in the countries in the Middle East where violence has erupted. We are announcing as well the second session of the third post-graduate course in geriatrics taking place in Dubai between 6-9th April 2011 for further information check the website <http://www/meama.com>

This issue has papers covering various topics from different countries in the region and from Ireland.

A case control study involving 90 elderly looked at the quality of life and functional level in Egyptian elderly on chronic hemodialysis treatment. The authors pinpoint that the proportion of older patients undergoing hemodialysis (HD) is rapidly increasing. The authors concluded that chronic kidney disease has a highly significant negative impact on quality of life of elderly patients and on functional decline.

A paper from Ireland reviewed Atrial Fibrillation (A.F) in the elderly which is the most common arrhythmia, affecting nearly 1% of the elderly population. The author stressed that the prevalence of AF among persons 60 years or older is 3.8%. whereas the prevalence of AF among persons 80 years or older is 10%. The author reviewed the current available management for AF.

A paper from Saudi Arabia presented a case of neuroleptic malignant syndrome associated with FAHRS Syndrome. The author mentioned that Fahr's disease (FD) or Bilateral Striato-PallidoDentate Calcinosi (BSPDC) which is a neurological entity characterized clinically by Parkinsonism, chorea, dystonia, ataxia, mental deterioration, seizures and neuropsychiatric features. The present case of ours is a rare presentation of Fahr's disease (Dandy walker malformation with Intra-cranial Calcinosi) with neuroleptic malignant syndrome in young girl who was on anti psychotics for long time.

Sandra Janet Ochoa from the State presented a nice analysis of the reason why to become a Clinical Analyst... Any Guidelines on Why I Should Be MD, MD/PhD or PhD?

A retrospective study from Jordan looked at the percentage of Keratoconus in Vernal Catarrh Patients. The study showed that Vernal catarrh was found to be associated with keratoconus. vigorous and frequent eye rubbing may aggravate or even cause keratoconus. The increased incidence of keratoconus was associated with male, long-standing disease, mixed and limbal forms, and advanced corneal lesions.

A retrospective analysis of Elderly Patients attending ENT Clinic with Ear Complaint at King Hussein Medical Center. A total of 160 patients aged 65 years or older were studied. The authors found that age-related hearing loss (presbycusis), wax in external auditory canal and tinnitus are the most common ear diseases encountered in elderly patients.

## **Study of QOL and functional level in Egyptian elderly, on chronic hemodialysis treatment**

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### **ABSTRACT**

**Background and objective:** The proportion of older patients undergoing hemodialysis (HD) is rapidly increasing. Individual experience largely drives advice on the prospect of life supported by renal replacement therapy and care for older patients supported by hemodialysis because there is a paucity of evidence. The objective of this study is to study the quality of life in elderly patients suffering from renal failure whether on conservative treatment or chronic hemodialysis regarding functional status, self perception and health related quality of life.

**Methods:** Case-control study. 90 elderly > 60years, 30 suffering from ESRD on hemodialysis, 30 renal failure on conservative therapy, 30 not suffering from renal disease from inpatient wards and outpatient clinics of Ain Shams University. Comprehensive Geriatric Assessment, SF36, ADL, IADL, and Karnofsky tools were performed. Statistical analysis (SPSS 17).

**Results:** Elderly on HD were more dependent in bathing, shopping, food preparation and house keeping. Scores of physical function, role limitation due to physical health, role limitation due to emotional problem and Karnofsky scale least in HD patients.

**Conclusion:** Chronic kidney disease has a highly significant negative impact on quality of life of elderly patients and on functional decline.

**Key words:** Hemodialysis, SF36, Karnofsky scale, ADL, IADL

## Introduction

The Kidney Disease Outcomes Quality Initiative (K/DOQI) of the National Kidney Foundation (NKF) defines chronic kidney disease as either kidney damage or a decreased kidney glomerular filtration rate (GFR) of less than 60 mL/min/1.73 m<sup>2</sup> for 3 or more months (1). Kidney disease is a growing problem in Egypt associated with an increase in the prevalence of dialysis patients. The statistics show a sharp rise in dialysis patients (2).

As physicians encounter a growing number of older patients, many of whom have multiple comorbid conditions, they will need to know how to look beyond the patient's physical health to the social environment and relationships that contribute to overall well-being and quality of life (3).

Functional status, an important domain of QOL refers to the ability to perform daily activities required to meet basic self-care needs and to maintain health and well being. Functional status reflects both functional capacity, what an individual is capable of doing, and functional performance, what an individual actually does in daily life (4).

Both HRQOL and functional status are strong predictors of adverse outcomes among patients with incident and prevalent End Stage Kidney Disease (ESKD) (5). Estimates of prognosis facilitate informed decision making and advanced care planning (6).

Patients' perception of their well-being and patient-reported outcomes (PROs) are becoming an integral part of evaluations of the human cost of chronic illnesses and the assessment of the impact of therapeutic interventions. Measures of health-related quality of life (HRQOL) have not only become popular investigative tools, but have been used in an effort to define and alter models of health care delivery. To date, there have been very few large-scale studies that have investigated the determinants of HRQOL in chronic kidney disease (CKD) patients not on dialysis. Most studies have tended to be small in size and cross-sectional.

Given the well documented, high mortality and hospitalization rate in CKD patients, understanding the HRQOL issues of CKD patients would seem to be an important area to explore (7). Moreover, the Egyptian elderly society with its diverse socioeconomic status and as a representative of an Arab, and yet developing country, needs to discover how renal disease has affected this increasing population.

"Adding life to years and not just years to life" is as true for ESRD patients as for any other individual (8).

## Methods

### Design:

The study is a case-control study.

### Sample:

90 elderly > 60 years

**1- Group I:** 30 elderly not suffering from renal disease.

**2- Group II:** 30 elderly with renal failure under conservative treatment.

**3- Group III:** 30 elderly with end stage renal disease under chronic hemodialysis.

All samples were recruited from the inpatient ward and outpatient clinics of Ain Shams University Hospital.

### Exclusion criteria:

1- Elderly suffering from depression.

2- Functional decline due to neurological or musculoskeletal diseases.

### Methods:

Every subject will undergo:

1- Comprehensive geriatric assessment, including:

2- Assessment of function using activity of daily living (ADL) and instrumental activity of daily living (IADL) (9).

3- Assessment of health-related quality of life by the short form 36 health survey (SF36) (10).

4- Assessment of quality of life regarding self perception using Karnofsky scale (11).

## Results

The only demographic data that was significantly different among the three groups was financial, where group II and III were significantly more financially dependent than group I. There was no significant difference between the different study groups as regards to different activity of daily living (ADL) except bathing; group III were more assisted than groups I and II.

Studying instrumental activities of daily living (IADL) revealed no significant difference between the different study groups as regards usage of telephone, laundry, medication intake, and financial handling. On the other hand shopping, food preparation and house keeping were not performed independently in group III.

As for the results of the Karnofsky scale the scores for group III were the lowest.

## Discussion

Studies on the quality of life of dialysis patients have not yielded consistent results with respect to the impact of disease and treatment. Quality of life (QOL) is an important parameter of the adequacy of treatment in dialysis patients (12).

We tried to avoid any significant difference between different study groups as regards demographic characteristics of different groups such as age, gender, marital status, work, and the special habits of medical importance as a number of demographic variables affect HRQOL. Among demographic factors, Seica et al, 2009 found older age, female gender, low socio-economic status and high educational level to be associated with lower QOL scores (8).

Hemodialysis patients undergo dialysis for four hours in a hospital per session of dialysis; that means they are away from their homes approximately three times per week for several hours (13). This would have a definite effect on their career plans, employment status, financial situation, as we found in the results that those patients are financially more dependent than the control group. Also there is effect on self-esteem and level of independence (14), which agrees with our results shown by patients with end stage chronic kidney disease undergoing hemodialysis.

Overall, the prevalence of functional impairment is high in elderly dialysis patients (15). Of particular concern is a lack of independent mobility, which is associated with a higher death rate (16).

	Group I	Group II	Group III	z <sup>#</sup> /p I-II	z <sup>#</sup> /p I-III	z <sup>#</sup> /p II-III
<b>Physical function</b>	80 (58-91)	63 (34-80)	25 (14-51)	-2.448	-4.772	-4.772
				<b>0.014†</b>	<b>&lt;0.001†</b>	<b>&lt;0.001†</b>
<b>Role limitation due to physical health</b>	88 (0-100)	0 (0-31)	0 (0-0)	-3.975	-4.565	-4.565
				<b>&lt;0.001†</b>	<b>&lt;0.001†</b>	<b>&lt;0.001†</b>
<b>Role limitation due to emotional problem</b>	100 (33-100)	33 (0-100)	17 (0-67)	-2.592	-3.442	-3.442
				<b>0.010†</b>	<b>&lt;0.001†</b>	<b>&lt;0.001†</b>
<b>Energy/ Fatigue</b>	55 (45-80)	50 (29-61)	35 (20-51)	-1.610	-3.842	-3.842
				0.107	<b>&lt;0.001†</b>	<b>&lt;0.001†</b>
<b>Emotional wellbeing</b>	66 (52-88)	62 (44-88)	68 (43-76)	-0.645	-0.682	-0.682
				0.519	0.496	0.496
<b>Social functioning</b>	94 (50-100)	63 (19-100)	0 (0-41)	-1.508	-4.872	-4.872
				0.132	<b>&lt;0.001†</b>	<b>&lt;0.001†</b>
<b>Pain</b>	68 (43-100)	45 (13-70)	28 (8-55)	-2.356	-3.756	-3.756
				<b>0.018†</b>	<b>&lt;0.001†</b>	<b>&lt;0.001†</b>
<b>General health</b>	55 (44-80)	53 (40-61)	38 (15-56)	-1.366	-3.041	-3.041
				0.172	<b>0.002†</b>	<b>.002†</b>

(Median, 1st-3rd Inter-quartile range)

†Significant/ z=z-value/p=p-value

Scores of physical function, role limitation due to physical health, role limitation due to emotional problem, and pain were least in group III.

Scores of energy/fatigue, social functioning, and general health were equally low in groups II& III.

**Table 1: Assessment of health-related quality of life using SF 36 scale**

When assessing functional status in this study there were more assisted cases in bathing in patients in group III than groups I and II but no significant difference between different study groups as regards to the other different activities of daily living (ADL), as shown by Francisco et al, 2008 in an epidemiological study on chronic renal failure elderly patients on hemodialysis, in which functional assessment assessed by means of the Katz index, found that the lowest independence score was in bathing, where 66.6% of included patients were independent for the item "bathing themselves" (17).

When studying IADL (Instrumental Activity of Daily Living), patients on chronic hemodialysis were more dependent in shopping,

food preparation and house keeping than control and CKD cases in conservative treatment groups. Subjects in the control group were more independent in usage of transportation means than subjects on chronic hemodialysis and CKD cases on conservative treatment. These results agree with the Tamura et al, 2009 study that concluded: the initiation of dialysis is associated with a substantial and sustained decline in functional status (18).

When assessing QOL using SF36 questionnaire the results showed a significant difference between the two case groups, in the same domains, where the CKD patients on conservative treatment scored higher than those on hemodialysis. The domains affected were

physical function, role limitation due to physical health, role limitation due to emotional problem, and pain. These results agree with numerous studies that prove that chronic hemodialysis showed significant affection of QOL.

A study by Kusumoto et al, 2008 showed that patients with terminal chronic renal insufficiency on hemodialysis often complain about a lack of energy, a feeling of discouragement and fatigue (19).

Chiang et al, 2004 study, which was a study for assessment of HR-QOL using the 36-Item Short-Form (SF-36) questionnaire, showed that the physical and mental aspects of quality of life are substantially lower for Taiwanese HD patients, except for higher bodily pain tolerance (20).

Also in the study done by Noshad et al, 2009, showed that the patients on PD had a better quality of life compared to those on HD in the following domains: psychophysical dysfunction, stress, sleep disorders and social dysfunction. In this study, technique, patients' survival and their quality of life were better on PD than on HD (14).

All of the above results can be explained by the fact that ESRD is a progressive, debilitating, chronic illness that requires nursing and medical interventions that include dialysis, education on lifestyle alterations and dietary and fluid restrictions. The disease also affects body image because of oedema and the presence of arteriovenous fistulae or a central venous catheter. The disease can have an impact on patients' QOL, potentially affecting their physical and mental health, functional status, independence, general well-being, personal relationships and social functioning (21). Traveling to an HD unit can be extremely time-consuming, especially if independent driving is no longer an option. Furthermore, patients who depend on public or ambulance transportation rather than a car have poorer quality-of-life measures and survival (22). The limitations of patients on hemodialysis, mainly physical, tend to increase with age, since the elderly have fragility due to the process of aging and are more prone to the occurrence of multiple comorbidities (23).

As regards the scores of Karnofsky scale, this study agrees with Ifudu et al, 1994 study which showed that maintenance hemodialysis does not return elderly patients to their predialysis level of functioning, it also found that the present functional activity had deteriorated compared with patients' score 2 years before initiation of hemodialysis (24).

Rebollo et al, 1998 study found that there was good HRQOL results of elderly transplant patients, in comparison with hemodialysis patients (25), also as found in Francisco et al, 2008 where Karnofsky performance scale scored less than 70, was found in 59.4% of the patients (17).

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(continued page 20)

## Atrial Fibrillation

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Atrial Fibrillation (A.F) is the most common arrhythmia, affecting nearly 1% of the elderly population. In the United States alone, 350,000 hospitalizations can be attributed to AF annually. The prevalence of fibrillation increases almost exponentially with age.

- The prevalence of AF among persons younger than 55 years is 0.1%.
- The prevalence of AF among persons 60 years or older is 3.8%.
- The prevalence of AF among persons 80 years or older is 10%.

In addition, incidence of AF continues to grow emphasizing the importance of identifying appropriate treatments that are safe, effective, and associated with improved clinical outcomes. A.FIB is classified as paroxysmal if it terminates spontaneously in fewer than 7 days (often in <24 hours). Atrial fibrillation is considered persistent when it terminates either spontaneously after 7 days or following cardio version. Atrial fibrillation that persists for more than one year, either because cardio version has failed or because cardio version has not been attempted, is called permanent atrial fibrillation.

- **Atrial fibrillation may cause symptoms such as**
- **Palpitations,**
- **Dizziness,**
- **Anxiety,**
- **Generalized weakness**
- **Mild shortness of breath.**

However, up to 90% of atrial fibrillation episodes may not cause symptoms. More serious signs and symptoms, such as chest pain, severe shortness of breath, and hemodynamic instability, may be due to associated cardiac disease such as ischemic heart disease or heart failure. Immediate cardio version is recommended in patients with instability due to atrial fibrillation.

Therefore, it is important to determine whether hemodynamic instability is due to the atrial fibrillation or other associated conditions.

When A.fib is suspected during auscultation of the heart with irregularly irregular beats, obtaining a 12-lead electrocardiography (ECG) is the next step. At a rate of 350-600 BPM the F waves may be seen as fibrillatory waves or may be absent. Unless the heart is under excess sympathetic or parasympathetic stimulation, the ventricular rate is usually between 80 and 180 bpm. With an abnormality in the intraventricular conduction system, the QRS complexes may become wide. It is important to pay attention to the electrocardiographic signs of associated cardiac diseases, such as left ventricular hypertrophy (LVH) and pre-excitation.

What is the role of electrocardiography in the diagnosis of A.FIB?  
When atrial fibrillation is suspected during auscultation of the heart

with irregularly irregular beats, obtaining a 12-lead electrocardiography (ECG) is the next step. Because atrial fibrillation is due to irregular atrial activation at the rate of 350-600 bpm with irregular conduction through the atrioventricular (AV) node, it appears on ECG as irregularly irregular narrow complex tachycardia. The F waves may be seen as fibrillatory waves or may be absent. Unless the heart is under excess sympathetic or parasympathetic stimulation, the ventricular rate is usually between 80 and 180 bpm. With an abnormality in the intraventricular conduction system, the QRS complexes may become wide. It is important to pay attention to the electrocardiographic signs of associated cardiac diseases, such as left ventricular hypertrophy (LVH) and pre-excitation.

### **Diseases which Predispose to A.FIB.**

Various cardiac diseases, including ischemic heart disease, valvular diseases, and cardiomyopathy, are associated with A.fib. Therefore, after the diagnosis of A.fib is confirmed with ECG or other cardiac tests, an evaluation of serum cardiac biomarkers and B-type natriuretic peptide (BNP) is usually required to investigate the underlying heart disease. More invasive cardiac tests (e.g., cardiac catheterization) may be required depending on the signs and symptoms and findings on the initial tests.

- Long-standing hypertension
- Ischemic heart disease
- CHF
- Any form of carditis
- Cardiomyopathy
- Infiltrative heart disease of any type
- Sick sinus syndrome
- Hyperthyroidism
- Pneumonia
- Lung cancer
- Idiopathic: Lone atrial fibrillation is idiopathic and defined as the absence of any known etiologic factors plus normal ventricular function by echocardiography.

Accordingly, chest radiography, serum thyroid-stimulating hormone (TSH) evaluation, CBC count, and serum chemistry are helpful, and other tests should be performed depending on the patient's presentation. Patients who are hemodynamically unstable, who have severe dyspnea or chest pain with A.fib, or who have pre-excited A.fib should undergo urgent cardio version. In stable patients with newly onset A.fib, the rate-control strategy should be attempted first to control the ventricular rate. If rate-control treatment does not elicit a response or if echocardiography does not reveal any valvular or functional abnormality of the heart, cardio version is indicated.

## TREATMENT OF A.FIB

### Medical cardioversion

Three main groups of drugs delay AV nodal conduction and are used for ventricular rate control. These include beta-blockers, nondihydropyridine, calcium channel blockers, and digoxin. Among these drugs, calcium blocker has become a popular initial choice because of its rapid onset of action and fewer negative effects. Beta-blockers and calcium channel blockers may reduce the blood pressure and cardiac function. Therefore, in atrial fibrillation accompanied by acute heart failure or hypotension, these agents may not be used; instead, digoxin is the choice for control in such cases. However, digoxin has a delayed onset of action so amiodarone is recommended for rapid pharmacological treatment of fibrillation accompanied by acute heart failure or hypotension. Calcium channel blockers and digoxin are contraindicated in fibrillation associated with pre-excitation; flecainide or amiodarone can be used. The Canadian Trial of Fibrillation (CTAF) and Sotalol Amiodarone Atrial Fibrillation Efficacy Trial (SAFE-T) studies showed that amiodarone is also more effective at maintaining sinus rhythm than other drugs. Propafenone has been prescribed in younger patients with lone paroxysmal symptomatic A. fibrillation. A beta blocker or calcium channel blocker should be used routinely or at least 30 minutes before propafenone, or other class drugs are taken to reduce the risk of the conversion of fibrillation to atrial flutter with 1:1 conduction.

### Which patient is considered unstable?

For example, if the patient who has dyspnea with mildly low or borderline oxygen saturation secondary to (COPD) exacerbation and has atrial fibrillation with a ventricular rate of 110 bpm, treatment of the COPD exacerbation (rather than cardio version to treat the atrial fibrillation) would be more beneficial. The heart rate of 110 bpm cannot explain the dyspnea and low oxygen saturation, and the patient's signs and symptoms are likely better explained by another cause.

### Electrical cardioversion

DC cardio version is the delivery of electrical current that is synchronized to the QRS complexes; it can be delivered in monophasic or biphasic waveforms. The required energy is usually 100-200 J (sometimes higher energy is required) for monophasic waveforms and less for biphasic waveforms. The patient should be sedated. In patients with A.fib of relatively short duration in whom the left atrium is not significantly large, the success rate of cardio version exceeds 75% (i.e., the size of the left atrium and the duration of A.fib inversely correlate with the success rate of cardio version).

### Complications of electrical cardio version

Embolization is the most important complication of cardio version. Accordingly, thrombus in the heart should be ruled out with transesophageal echocardiography or warfarin should be given for anticoagulation for 4 weeks before cardio version is performed. In addition, the patient should receive anticoagulants for at least 4

weeks following the procedure. Complication of electrical cardio version may include pulmonary edema, hypotension, myocardial dysfunction, and skin burns, which may be avoided with the use of steroid cream and proper technique. Electrical cardio version is also associated with some ST- and T-wave changes on ECG.

### Who should undergo urgent cardio version?

Urgent cardio version is indicated in unstable patients with newly onset A.fib, if the duration of atrial fibrillation exceeds 48 hours or is unknown, heparin should be administered concurrently. For 4 weeks following the procedure, warfarin should be administered for anticoagulation, and the target international normalized ratio (INR) should be between 2 and 3. Limited data support the use of low molecular heparin.

### Role of transesophageal echocardiography and anticoagulation therapy before cardioversion

The American cardiology centre strongly recommends anticoagulation (INR between 2 to 3) be administered at least 3 weeks prior to and 4 weeks following cardio version in patients with A.fib of 48 hours' duration or longer or of unknown duration, regardless of the method. In this situation, as an alternative to anticoagulation prior to cardio version, it is reasonable to perform TEE to search for thrombus in the left atrium or left atrial appendage. In patients with no identifiable thrombus, immediate cardio version with concurrent heparinization and subsequent anticoagulation for 4 weeks is an option. However, patients in whom TEE reveals thrombus should receive anticoagulation for at least 3 weeks prior to and 4 weeks following cardio version.

Follow-up Investigation of Rhythm Management (AFFIRM) study showed that the rate control or sinus rhythm yields similar results in terms of mortality rate and stroke rate. Rate control has become a popular strategy in most patients. This is particularly true in elderly patients and in those who would benefit from rate-control drugs because of other associated diseases (e.g., other cardiac diseases).

### Rate control should be assessed both at rest and with exertion

It is advisable to refer the patient to a cardiologist for initiation of an anti arrhythmic drug unless the physician is experienced with and knowledgeable of administration of these drugs. The initiation of anti arrhythmic drugs is associated with uncommon but life-threatening arrhythmic adverse effects; accordingly, an initial 24 hours of in-hospital monitoring is generally advised. However, the ACC recommends outpatient initiation of propafenone or flecainide therapy in patients with lone paroxysmal fibrillation and no structural heart disease who are in sinus rhythm at the time of drug initiation. The ACC (American Cardiological Centre) also recommends outpatient administration of sotalol in patients who are in sinus rhythm and who have little or no heart disease, are prone to paroxysmal fibrillation with a baseline uncorrected QT interval of fewer than 460 milliseconds, have normal levels of serum electrolytes, and have no risk factors for class III drug-related pro-arrhythmia.

Rate-Controlling Agents	Adverse Effects
Amiodarone	Hypotension, heart block (HB), pulmonary toxicity, skin discolouration, hypothyroidism, hyperthyroidism, corneal deposits, optic neuropathy, warfarin interaction, sinus bradycardia
Digoxin	Digitalis toxicity, HB, bradycardia
Esmolol	Hypotension, HB, asthma, heart failure (HF)
Metoprolol	Hypotension, HB, bradycardia, asthma, HF
Propranolol	Hypotension, HB, bradycardia, asthma, HF
Diltiazem	Hypotension, HB, HF
Verapamil	Hypotension, HB, HF

## Percentage of Keratoconus in Vernal Catarrh Patients in South of Jordan

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### ABSTRACT

**Aims:** To detect the incidence of keratoconus in patients with vernal catarrh.

**Methods:** A retrospective study of 45 patients (90 eyes) with vernal catarrh, age group extending from 11 to 23 years, attending the ophthalmic clinic in Prince Zaid bin Al-Hussein Hospital in South of Jordan from January, 2007 to March, 2008. All participants visited the clinic for complete history and an eye examination (slit-lamp biomicroscopy, retinoscopy and keratometry).

**Results:** The study included 30 male and 15 female patients (90 eyes). The clinical types of vernal catarrh were as follows: 42.2% mixed, 33.3% palpebral and 24.4% limbal types. Twenty-two eyes (24.4%) of 90 eyes were detected as keratoconus. Thirty-four eyes (37.7%) had keratopathy as a complication of vernal catarrh such as pseudogerontoxon, punctate keratitis, and corneal ulcer.

**Conclusions:** Vernal catarrh was found to be associated with keratoconus; vigorous and frequent eye rubbing may aggravate or even cause keratoconus. The increased incidence of keratoconus was associated with male, long-standing disease, mixed and limbal forms, and advanced corneal lesions.

### Introduction

Keratoconus is defined as a noninflammatory condition characterized by the change in eye form from normal round shape to a cone shape. Which results when the cornea thins and protrudes (results from the bilateral central or axial ectasia of the cornea).

The etiology of keratoconus is unknown. Keratoconus has been associated with atopy (1,2) atopic dermatitis, vernal catarrh(3), Downs syndrome, retinitis pigmentosa, Marfans syndrome, aniridia, blue sclera, and mitral valve prolapse.

**Clinical Presentation:** Keratoconus often manifests during the late teens or early twenties, then progresses slowly for the next decade or two as the cornea scars and becomes more elongated. Myopia and an irregular astigmatism may result from the alteration in the normal corneal contour.

**Treatment:** Some authors recommend cautioning patients about rubbing their eyelid as some believe this hastens progression of keratoconus. Mild cases of keratoconus can be treated with glasses or contact lenses. Often a rigid lens compensates for the irregular corneal astigmatism. Some have tried superficial keratectomy, epikeratoplasty and thermokeratoplasty(4) with variable and often temporary improvement. Penetrating keratoplasty(5) is considered very successful for long term rehabilitation in severe cases. A new treatment option for keratoconus is Implantation of intracorneal ring segments with support from the femtosecond laser(6).

Vernal catarrh is a chronic, bilateral conjunctival inflammation, predominantly observed in children and young adults(7). It occurs more commonly in males than females, with an onset typically between 8 and 10 years of age with photophobia and intense itching that recurs seasonally during warm weather, prevalent in areas

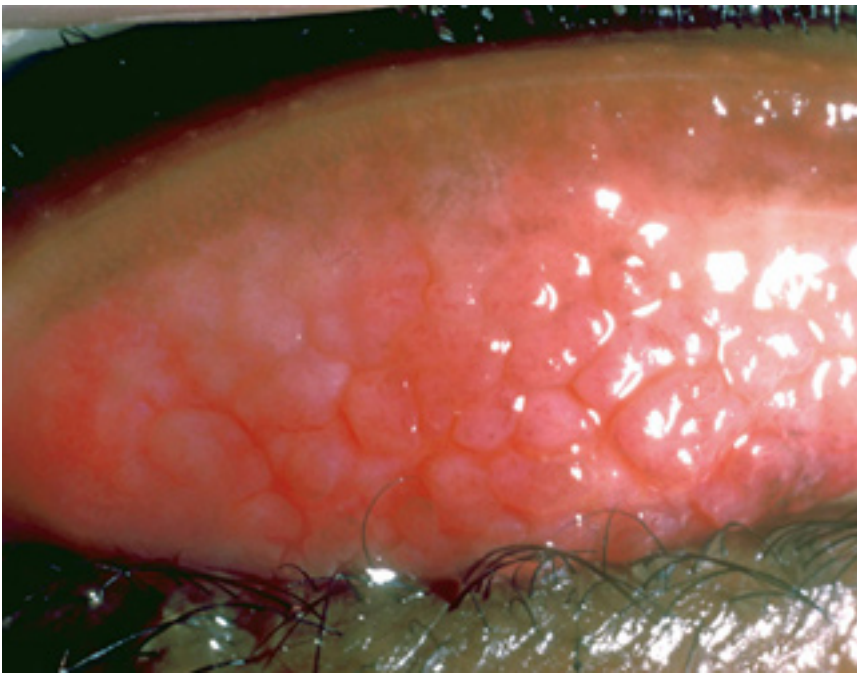


Figure 1: Giant papillary hypertrophy with the typical cobblestone appearance of the upper tarsal conjunctiva

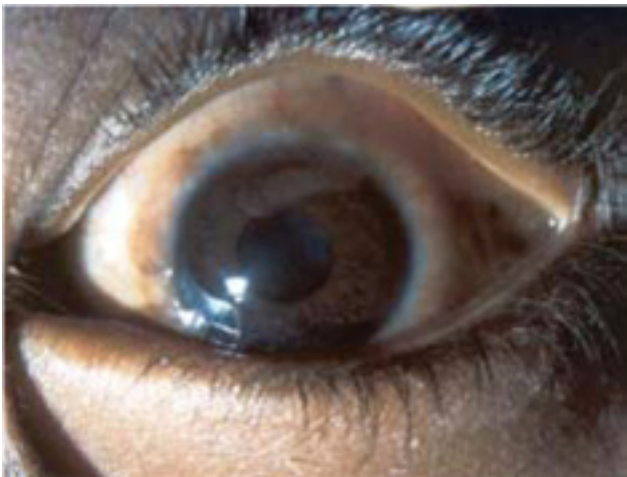


Figure 2: Limbitis with Horner- Trantas dot

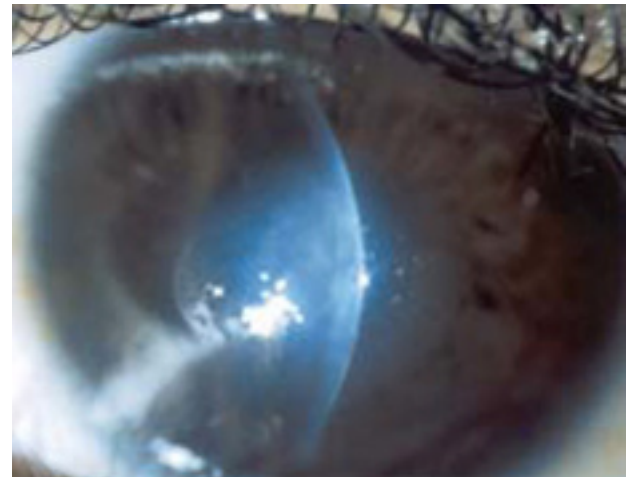


Figure 3: Acute corneal hydrops

which are hot and dry such as the Mediterranean area and the Middle East. The disease occurs in palpebral, limbal (limbitis see Figure 2) and mixed forms. Corneal involvement occurring in about 50% of the cases, is characterized in the palpebral form by cobblestone papillae in the upper palpebral conjunctiva (see Figure 1) and in the bulbar form by gelatinous nodules adjacent to the corneoscleral limbus(8)(Horner-Trantas dot) Figure 2. The corneal complications of vernal keratoconjunctivitis include(9) superficial punctate keratitis, corneal epithelial erosions (Macro erosions), sub epithelial scarring, corneaectasias (the commonest and most frequent being keratoconus), acute hydrops ( see Figure 3), conjunctivalisation of the cornea, vascularisation and pannus formation, shield ulcers(10) and corneal plaques, pseudogerontoxon, contact lens failure, sterile stromal melting and corneal perforation.

**Treatment of vernal catarrh includes the following**

1. Removal of allergen from environment
2. Astringent lotion, adrenalin 1:10000, antihistaminic drops (chlorpheniramine), mast cell stabilizers (sodium

- cromoglycate, olopatadine, ketotifen etc)
3. Short course corticosteroid drops
4. Topical 2% sodium cromoglycate drops(11).
5. Others: using topical cyclosporine for treating shield ulcers(12,13), cryotherapy, surgical or laser assisted excision of giant papillae(14) with or without mitomycin(15), excimer laser phototherapeutic keratectomy, amniotic membrane graft.

**Materials and Methods**

The diagnosis of vernal keratoconjunctivitis depended on typical signs and symptoms and was categorized into palpebral, limbal and mixed as follows:

<b>Palpebral</b>	papillae > 1 mm on tarsal conjunctiva with no infiltration
<b>Limbal</b>	Limbal gelatinous involvement of the limbus, limbitis - brownish hyperpigmentation around the limbus and/or Horner-Trantas dots
<b>Mixed</b>	both palpebral and limbal involvement

The cases were included as having a diagnosis of keratoconus where there was evidence of at least two of the following three clinical signs; central conical corneal protrusion, positive Munson's sign and central corneal thinning (unless there was previous hydrops).

Severity of keratoconus was assessed using the average of the two principal meridian keratometry (K) readings;

<b>Mild</b>	<48 D
<b>Moderate</b>	48-54 D
<b>Severe</b>	>54 D

A retrospective study of 45 patients with a mean age of 17 years (11years-23years) ; 30 patients were males, 15 patients were females, attending the ophthalmic clinic by taking a complete history and eye examination. Examination included the usual methods of retinoscopy, ophthalmoscopy, and biomicroscopy. Patients with keratoconus were also examined with keratoscopes. Intraocular pressures were usually checked with applanation tonometer, and we report here our results on the incidence of keratoconus in the present series of patients with vernal catarrh.

## Results

Of the 45 participants (90 eyes), 35 males and 15 females were involved in this study. The clinical types of vernal catarrh were as follows: 42.22% mixed, 33.33% palpebral and 24.44% limbal types, (see Table 1). Signs of vernal catarrh were commoner in patients under the age of twenty years.

Twenty-two eyes (24.4%) of 90 eyes were detected as keratoconus. Thirty-four eyes (37.7%) had keratopathy as a complication of vernal catarrh such as pseudogerontoxon, punctate keratitis, and corneal

ulcer. Of the 32 eyes affected with keratoconus, 8 eyes were legally blind, with a visual acuity of 6/60 or less, while 3 eyes had marked visual handicap with a vision of more than 6/60 but less than 6/24 (see Table 2).

Bilateral keratoconus occurred in the majority of cases (75 eyes) nearly 83%.

The assessment of severity of keratoconus depended on the keratometry readings and these measurements could not be obtained in three patients (six eyes), one of whom was mentally retarded children. Five eyes had received corneal grafts. Of the remaining 16 eyes, 8 eyes (50%) showed severe keratoconus, 3 eyes (18.75%) moderate and 5 eyes (31.25%) showed mild keratoconus (see Table 3 next page).

## Discussion

Vernal keratoconjunctivitis is a common form of ocular allergy especially in tropical countries. The disease was found to lead to significant changes in the ocular surface often causing severe visual impairment in one or both eyes.

All patients of keratoconus in this study were suffering from vernal catarrh.

Keratoconus has been associated with a familial pattern of inheritance. Both dominant and recessive traits have been reported; consanguinity has also been blamed in some such cases. In our study we did not find any history of keratoconus among family members. This leaves us only with the theory of eye rubbing secondary to the intense irritation caused by the severe mixed type of vernal catarrh. In our patients, eye rubbing was strongly and significantly associated with vernal, as might be expected, and might have played a role in the development of keratoconus in those patients.

Clinical Types of Vernal Catarrh	No. of eyes	%
<b>Palpebral</b>	<b>30</b>	<b>33.33</b>
<b>Limbal</b>	<b>22</b>	<b>24.44</b>
<b>Mixed</b>	<b>38</b>	<b>42.22</b>
<b>Total</b>	<b>90</b>	<b>100</b>

Table 1: The clinical types of vernal catarrh

Visual status	No. of eyes	%
<b>HM-6/60)</b>	<b>8</b>	<b>36.36</b>
<b>6/60-6/24</b>	<b>3</b>	<b>13.63</b>
<b>6/18</b>	<b>5</b>	<b>22.72</b>
<b>6/12</b>	<b>2</b>	<b>9.09</b>
<b>6/9</b>	<b>1</b>	<b>4.54</b>
<b>6/6 (after PK*)</b>	<b>3</b>	<b>13.63</b>
<b>Total</b>	<b>22</b>	<b>100</b>

Table 2 : Best correctable visual acuity in eyes with keratoconus

Severity of keratoconus	the keratometry readings	Number of eyes	%
Mild	<48 D	5	31.25%
Moderate	48-54 D	3	18.75%
Severe	>54 D	8	50%
Total		16	100%

Table 3 : Severity of keratoconus

So we conclude from our study that there is a relationship between the keratoconus, vernal catarrh (vernal keratoconjunctivitis) and eye rubbing. This triad is found to be responsible for the increase in the incidence of keratoconus in the South of Jordan especially Tafila because the weather in these regions has a role in developing the allergic type of keratoconjunctivitis (hot, dryness, dust, beside the presence of animals, trees (pollens), flowers, etc

Also we conclude that the increased incidence of keratoconus was associated with male, long-standing disease, mixed and limbal forms, and advanced corneal lesions.

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## Elderly Patients attending ENT Clinic with Ear Complaint

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### ABSTRACT

**Objective:** To carry out a retrospective analysis of the ear diseases in elderly patients attending Ear, Nose and Throat clinic at King Hussein Medical Center.

**Patients and Methods:** A total of 160 patients aged 65 years or older attending the department of Oto-Rhinolaryngology, Head and Neck Surgery of King Hussein medical center (Amman- Jordan), between March 2006 and January 2011 with a chief complaint of ear disease were taken up.

**Exclusion criteria** were those patients below the age of 65 and those attending the clinic with no ear complaint.

The following data were recorded for each patient: age, sex, history of otological disease; duration of complaint and final otological diagnosis.

**Results:** The total number of patients was 160 and it comprised 84 men (52.5%) and 76 women (47.5%). Ages ranged from 65 to 84 years, with mean age of 67.3. The majority of patients fall within the age group of 65-70 years (57.7%) while the least (3.1%) fall in the age group 81-85 years. The most common ear disease was age-related hearing loss, presbycusis, 55 (34.4%) which was followed by wax in external auditory canal, 36 (22.5%). Tinnitus of undetermined etiology was a common reason for a hospital visit, 23 (14.4%). Among patients with infective ear disease, infection of external canal (otomycosis and acute otitis externa) was the commonest. Other diseases encountered in these patients were chronic suppurative otitis media, acute suppurative otitis media, and vertigo and foreign bodies in external auditory canal.

**Conclusion:** Age-related hearing loss (presbycusis), wax in external auditory canal and tinnitus are the most common ear diseases encountered in elderly patients. Full audiological and otological evaluation of these patients should be carried out on regular bases.

**Key words:** Elderly, Ear diseases, Presbycusis

### Introduction

Care of the elderly patient involves some fundamental premises which must be taken into account in treatment by otolaryngologists. Often multiple diseases coexist in these patients which often present a diagnostic dilemma in treatment. The elderly also suffer from a unique set of illnesses which only occur in old age. Illnesses can present with unusual symptoms without common symptoms of pain and fever which may lead to diagnostic dilemmas. Otolaryngologists play a pivotal role in prevention of illness and other problems in the head and neck area.

The aging process in humans affects all the organs of the body, including the ear. It has been noticed an increase of elderly people with hearing disorder-related symptoms due to the rising life expectancy of this population. Complaint of "listen, but do not understand" symptom, mainly in a noisy environment, where there is music and people talking, is becoming more frequent from people aged over 60.

One of the greatest challenges in the geriatric population is their ability to communicate their problems, needs, and desires in a medical setting. Hearing impairments can hamper a patient's ability to express himself or herself clearly or to understand questions or

commands. This is an enormous burden on the patient, the caregiver, and the psychiatrist as they work to achieve rehabilitation goals. Addressing these issues on the patient's initial visit can ameliorate problems and prevent frustration and further difficulties.

The aim of the present study is to analyze ear diseases in elderly patients attending the Ear, Nose and Throat clinic at King Hussein Medical Center.

## Patients and Methods

In this retrospective study, 160 patients aged 65 years or older attending the department of Oto-Rhinolaryngology, Head and Neck Surgery of King Hussein medical center (Amman- Jordan), between March 2006 and January 2011 with the chief complaint of ear disease, were taken up.

Exclusion criteria were those patients below the age of 65 and those attending the clinic with no ear complaint.

All cases underwent a detailed history taking and a thorough general examination, systemic examination and examination of the nose, throat and ears with special emphasis on identifying the cause of their ear complaint. Patients who presented with a complaint of decreased hearing were subjected to otoscopic examination and full audiological evaluation.

The following data were recorded for each patient: age, sex, history of otological disease; duration of complaint and final otological diagnosis.

## Results

The total number of patients was 160 and it comprised 84 men (52.5%) and 76 women (47.5%) Table I. The main presentation of these patients was ear complaint with a duration ranging from 3 days to 4 years.

Ages ranged from 65 to 84 years, with mean age of 67.3. The majority of patients fall within the age group of 65-70 years (57.7%) while the least (3.1%) fall in 81-85 years. Table II.

Most common ear disease was age-related hearing loss, presbycusis, 55 (34.4%) which was followed by wax in external auditory canal, 36 (22.5%). Tinnitus of undetermined etiology was a common reason for a hospital visit, 23 (14.4%). Among patients with infective ear disease, infection of external canal (otomycosis and acute otitis externa) was the commonest. The other ear diseases are shown in Table III.

## Discussion

Geriatric patients present 3.5 times more health problems compared to the young population (1,2). Health problems of geriatric people cause dependency and depression which when added to otorhinolaryngological problems, makes the social interaction of geriatric patients worse. The frequency of otorhinolaryngological diseases appears to start increasing around 40 years of age with an even steeper increase beginning around 60 years of age. Hearing impairment has been found to be the most common otorhinolaryngological morbidity (3)

Presbycusis is the most common form of hearing loss encountered in old age. It is the main disorder found in this series. This is as expected since presbycusis is a slowly progressive decline in threshold of hearing due to aging process. It is a complex disorder that in addition to loss of perception to pure tones involves loss of speech processing and discrimination.

Presbycusis had been consistently found in different populations. Its aetiology has numerous variables such as diet, nutrition, metabolism, cholesterol levels, blood pressure, arteriosclerosis, exercise, smoking, exposure to noise, emotional stress, and genetic factors.

The high prevalence of presbycusis in this study corresponds to the results obtained elsewhere in similar study populations (1,4).

World Health Organization (WHO), reports that 30 - 35% patients above 60 years suffer from presbycusis and this increases to 40 - 45% in patients above 70 years of age (5,6).

Rosenhall confirms the results of population based studies that had shown a higher prevalence of hearing loss in this age group (7). Cruickshanks et al found still higher prevalence of hearing loss (45.9%) in an epidemiological study of geriatrics in Wisconsin, USA (8). Hearing loss severely affects the quality of life especially in the background of low socioeconomic status where the accesses to health care facilities are restricted due to various reasons (9-13). It increases the disability burden on society and could be a cause of depression, isolation and suicidal tendencies.

Wax in the external auditory canal was the second most common finding in our study (22.5%). The quoted incidence of this was reported between 8% and 35% (14-16). Our result is similar to previous reported studies (15,16).

It is a reversible, frequently overlooked cause of conductive hearing loss in elderly people (15-18). There may be associated earache, sensation of ear blockage, and tinnitus. These symptoms are particularly distressing to elderly patients who may already have compromised hearing. Removal of wax significantly improves hearing ability and prevents serious social withdrawal due to hearing loss (15-18).

Wax removal must be carefully done by qualified personnel, because removal by an inexperienced person can cause damage to the external auditory meatus, traumatic perforation of the tympanic membrane, and/or otitis media (18,19).

Tinnitus of undetermined etiology was a common reason for a hospital visit in our study (14.4%).

Tinnitus is common among the elderly (20,21), and a substantial proportion of older people suffer from their tinnitus (22).

It has been suggested that older persons with tinnitus suffer more than their younger counterparts (22). One possible factor is hearing loss, which is one of the best-known precursors of tinnitus, and also increasingly more common in old age. In addition, aging is accompanied by a decline in several sensory and cognitive functions. Elderly people do not receive help for their health problems to the same extent as do younger people (23), and this is likely the case with tinnitus.

Infection is a significant cause of morbidity among our elderly patients. Of these, infection of external auditory canal (otomycosis and acute otitis externa) is distinctively the most common encountered in our study. Other causes include chronic and acute suppurative otitis media.

## Conclusion

Age-related hearing loss (presbycusis), wax in external auditory canal and tinnitus are the most common ear diseases encountered in elderly patients. Full audiological and otological evaluation of these patients should be carried out on regular bases.

Sex of patients	Total No.	Percentage
Male	84	52.5%
Female	76	47.5%
<b>Total</b>	<b>160</b>	<b>100%</b>

Table I: Distribution of patients with respect to sex (No. 160).

Age of presentation	Total No.	Percentage
65-70 years	92	57.5%
71-75 years	43	26.9%
76-80 years	20	12.5%
81-85 years	5	3.1%
<b>Total</b>	<b>160</b>	<b>100%</b>

Table II: Distribution of patients with respect to age (No. 160).

Clinical Diagnosis	Total No.	Percentage
Presbycusis	55	34.4%
Wax in external auditory canal	36	22.5%
Tinnitus (Idiopathic)	23	14.4%
Otomycosis	12	7.5%
Acute Otitis Externa	11	6.9%
Chronic suppurative Otitis Media	9	5.6%
Vertigo	7	4.4%
Foreign body in the external auditory canal	4	2.5%
Acute suppurative Otitis Media	3	1.8%
<b>Total</b>	<b>160</b>	<b>100%</b>

Table III: Clinical diagnosis of ear disease (No. 160).

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## I Would Like to Be a Clinical Analyst... Any Guidelines on Why I Should Be MD, MD/PhD or PhD?

Okay. .my advice is the fact that prior to when you decide to set yourself on one of these paths perform some clinical shadowing plus some lab research.

Some definitions first....

MD: Indicates Doctor of Medicine, a doctor's diploma in medicine

PhD: Is the highest qualification obtained at a college or university, usually requiring 3 to 5 years of original analysis in a particular field of study.

MD/PhD: refers to an education consisting of both the medical training of a doctor (MD or DO) with the rigor of a scientific analyst (PhD)

You could also like to try to get involved in some clinical research.

This will likely provide you a taste of the different fields. Some MDs do clinical research, if you get interested in that, you would not need an MD/PhD.

You actually should gain some quality exposure prior to making any decisions. Neither clinical work nor lab bench job is exactly what it might seem like in theory. You need to get your hands dirty. Attempt to request information, find out about them, and obtain a couple of tastes of each one.

I believe it's more easy to find a personality niche when you are content with the specific work you're doing daily, rather than make an effort to enjoy doing work you hate, even if you fit the "typical profile" of the job.

Generally a double degree is perfect for people who find themselves interested in both, basically. However, you will possibly not find yourself doing a lot of the actual bench work if you are an MD/PhD. The MD/PhD that is the P.I. of the laboratory I currently work for NEVER does one of the actual experiments we currently do; he simply manages administrative stuff and discusses problems/ideas together with his henchmen.

Most of his time in the week is spent on clinical work. I am not sure that may be the way it always works, but this can be my own experience. However, if you might be equally interested in both, then I would still think an MD/PhD will probably be worth considering.

MD/PhD will place you at some advantage in grant-writing while you're a new researcher. (Eventually, the degree matters less because research recruiters assess you depending on your actual accomplishments.)

Imagine that studying scientific research will be easier if you have been trained like a physician. This advantage isn't well worth the extra 3 years, but it's somewhat of an advantage. It provides you with the flexibility to determine patients if you want. A slight majority of the MD/PhD's I have seen usually do not, but some do and in any case all of them could. It will assist in the pursuit of an academic position too.

And also you? What are your benefits and drawbacks of selecting a MD, MD/PhD or PhD job?

Who am I ? Sandra Janet Ochoa is writing for the <http://www.clinicalresearchtraining.net/> clinical research training program blog, her own and non-commercial in nature hobby blog to deliver free suggestions for clinical research training newbie's/experts to assist them get a new career.

## Neuroleptic Malignant Syndrome Associated With Fahr's Syndrome

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### INTRODUCTION

Fahr's disease (FD) or Bilateral StriatoPallidoDentate Calcinosis (BSPDC) is a neurological entity characterized clinically by Parkinsonism, chorea, dystonia, ataxia, mental deterioration, seizures and neuropsychiatric features, and pathologically [1] by massive bilateral calcification of the : basal ganglia, sulcal depths of the cerebral cortex, dentate nuclei of the cerebellum, internal capsule and lateral parts of the thalamus. Common clinical features include movement disorders such as parkinsonism, speech disorders, psychiatric disorders, epileptic seizure, dementia, cerebellar or extra-pyramidal dysfunction.[2,3] . These cases have increased incidence of neuroleptic malignant syndrome (NMS). The present case of ours is a rare presentation of Fahr's disease (Dandy walker malformation with Intracranial Calcinosis) with neuroleptic malignant syndrome in young girl who was on anti psychotics for long time.

**Key words:** Movement Disorder , Intra-Cranial Calcification

### Case Report

This 16 year old female was referred to us from a specialty psychiatry hospital with a chronic history of difficulty in performing her routine physical activities of daily living associated with increasing slowness in her work of one week duration and for the last three days the patient's mother noticed that she was most of the time lying straight and stiff in the bed and had started running a high fever. She was clinically diagnosed as a case of bipolar affective disorder for the last 2 years and had been on a regular drug treatment for it, but lately she started having aggressive behavior since the past three months, and she was put on haloperidol 5mg t i d, along with depot preparation of fluphenazine 25mg once biweekly. She had a history of mental retardation since child hood and poor schooling. She is a product of a non consanguineous marriage and her other siblings are normal.

Examination on admission revealed that she was conscious, irritable, febrile with a recorded body temperature of 39.0 Celsius (axillary) with marked rigidity observed all over her body associated with tremors. The latter were clinically both resting as well as action tremors. She was dehydrated and her vital signs were: Pulse 116 beats per minute, regular; BP: 134/88 mmHg; Respiratory rate: 20 breaths per minute and a core body temperature of 40.4°C (rectal).

Investigations showed mild leukocytosis, normal renal parameters and liver functions. Her CPK was elevated at 1678 Units. Non contrast CT scan of the brain showed a midline CSF-filled posterior fossa cyst which is communicating anteriorly with a mildly dilated fourth ventricle but the posterior fossa typically is normal in size (Figure 1). CT scan of the supratentorial region showed diffuse calcifications of basal ganglia,

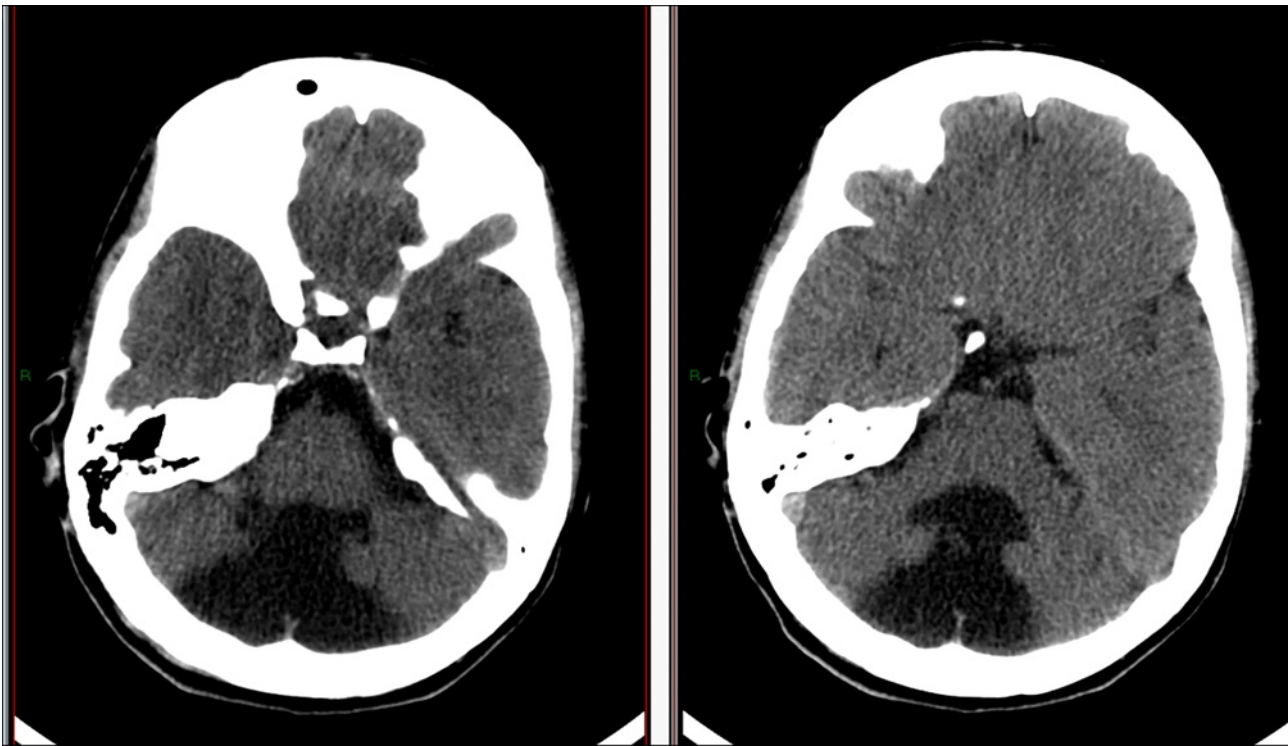


Figure 1

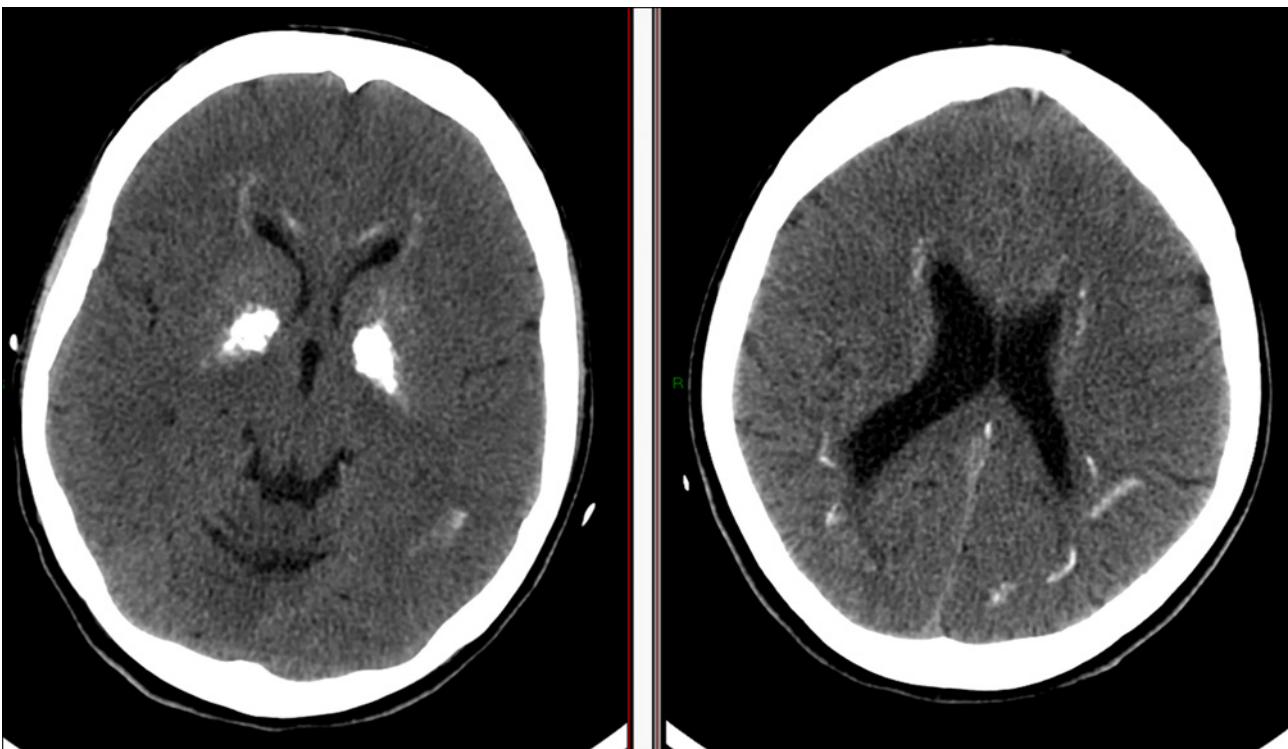
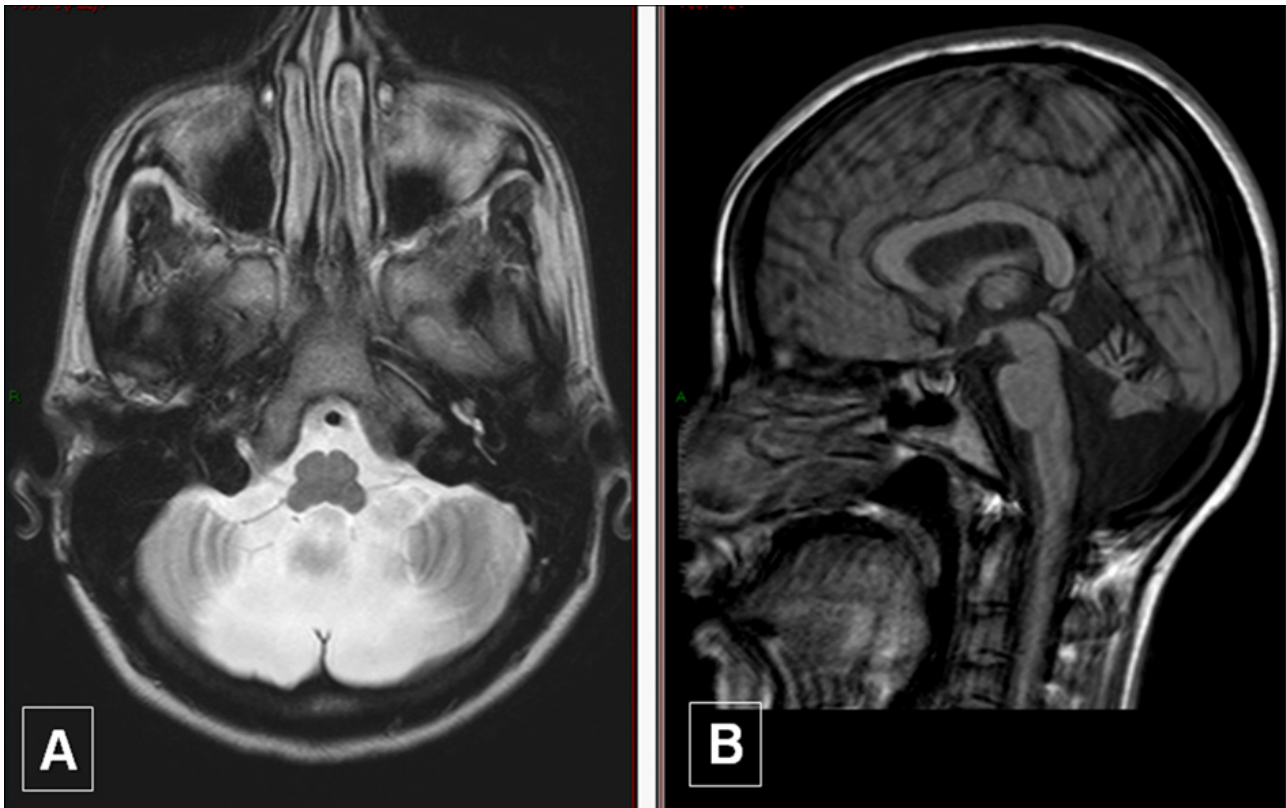


Figure 2

periventricular white matter, centrum semiovalae and cerebral subcortical regions (Figure 2). No associated brain edema or mass effect was seen. No abnormal enhancement was seen after contrast administration. MRI of the brain showed a mid-line cerebrospinal fluid cyst in the posterior fossa, hypoplastic cerebellar hemispheres and vermis. The cyst has winged appearance and it is communicating with the fourth ventricle (Figure 3A and B)

CSF analysis was done and was within normal limits; serum calcium of 9.6 mg/dl [(9-11) milligrams per deciliter (mg/dL)]

and serum phosphorus of 3.8mg/dl[2.4 - 4.1 milligrams per deciliter (mg/dL)]. Her serum Parathyroid hormone(PTH) level was normal [ 49 pg/ml (10-55 picograms per milliliter (pg/mL))] and her thyroid function was also normal. TORCH and Brucella serologies were both unremarkable. The patient was diagnosed as a case of neuroleptic malignant syndrome and she was immediately started on treatment with aggressive iv fluid hydration and bromocriptine together with the withdrawal of the anti-psychotics both the long acting phenothiazine(fluphenazine) and short-acting butyrophenone(haloperidol, the more implicative agent in the causation of NMS).



*Figures 3A and 3B*

Gradually she started improving and 2 weeks later was discharged in a clinically good condition and a neuropsychiatrically stable condition with advice on discharge to be on regular medical and psychiatric follow-up.

## Discussion

Intracranial calcification has been a finding in about 1% of patients who undergo brain CT.

One study of a psychiatric population, found calcification of basal ganglia in 0.087%, of a large series of patients of mixed diagnoses[3]. Lowenthal set forth pathological criteria defining FD, including characteristic pallidal calcification evident on CT and on macroscopic pathological exam[4]. Although clinical manifestations vary, clinical definitions have been devised. One definition requires bilateral calcifications with neuropsychiatric and extrapyramidal disorders attended by normal calcium and phosphorus metabolism. Another stipulates the occurrence of seizures, rigidity and dementia with a characteristic calcification of the basal ganglia. Neuropsychiatric symptoms, often the first or most prominent manifestations, range from mild difficulty with concentration and memory to changes in personality or behavior to psychosis and dementia. It has been suggested that those who become symptomatic early in adulthood are more likely to have psychosis. Symptomatic features may change over time. More extensive calcification and subarachnoid space dilatation are known to correlate with the presence of psychiatric manifestations (Konig et al, 1989)[5]. Our patient had extensive intracranial calcification with normal calcium and phosphorus metabolism with normal parathyroid function. Her psychiatric

symptoms were first to get manifested and that too at an early age.

Neuroleptic malignant syndrome is a fatal treatment related complication. Initial description was by Delay in French literature : “0.2% to 2% of patients on antipsychotics develop NMS”[6]. Since its occurrence is linked to a decrease in dopaminergic transmission it is reported more frequently with high potency antipsychotics such as haloperidol. Situations where NMS may occur are:

- 1. Initiation on a high dose of high potency antipsychotic or increase of antipsychotic dose ;**
- 2. Switch from a low potency to high potency antipsychotic, and**
- 3. Sudden withdrawal of dopaminergics in patients of Parkinson’s disease.**

In addition it occurs sometimes when antipsychotics are used for other reasons, such as for vomiting or sedation. In these situations NMS is often not suspected and hence diagnosis is missed. The potential morbidity of neuroleptic malignant syndrome in patients with striato-nigral degeneration is more. Our case had severe presentation of neuroleptic syndrome which can be correlated with co-existent basal ganglionic degeneration (calcification)[7].

Our patient had cerebellar atrophy along with Dandy walker malformation; this unusual combination has not been reported in the literature.

## Conclusion

The association of Fahr's disease with Neuroleptic Malignant Syndrome is a rare clinical presentation reported to be having an incidence of less than 0.5% in available medical literature. Our case is the first in this series to have been reported from this geographical region of western Saudi Arabia. This disorder needs to be considered on the basis of its clinical presentation in combination with initial CT-brain and later by a conclusive MR brain scan lest the diagnosis may be missed and the patient gets treatment for a wrong diagnosis as was the initial scenario in our patient.

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