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## Middle East Journal of Age and Ageing

# Contents

# Editorial

*Dr Abdul Abyad*
Chief Editor

This is the first issue this year and we are continuing our series of papers on ageing in different countries.

We are starting this year with a paper on ageing and security in Tehran. The paper investigates life security in the course of ageing in Tehran City which is generalizable to other parts of the country. The present survey studies various dimensions of the lives of the elderly people ___ their material quality of life, their wellbeing, their abuse etc. The author tries to reflect the quality of human rights and the dignity of the ageing people in the present paper. Though the number of the elderly people is increasing in Iran, yet institutions enough haven not been built to meet their increasing needs. The method of research used, is mainly empirical, yet it is preceded by theoretical and literature review. While five hundred elderly people have randomly been selected for the study, the main hypothesis of the study is: "increasing security of the elderly positively enhances their quality of life". The paper concludes that the new generation of the elderly with new needs and expectations are quite different from those of the previous generations and age cohorts.

A paper from Qatar in fulfillment of a thesis degree looked at Antibiotic Prophylaxis to prevent Urinary Tract Infection Following Urinary Catheterization in Geriatrics.

The author stressed that Urinary Catheterization is common procedure in elderly population. Literatures showed that up to 40% of the nosocomial infections are caused by urinary tract infection related to urinary catheterization. Some studies suggest using prophylaxis antibiotics prior to insertion of urinary catheter in elderly to minimize the chances of urinary tract infection related to the procedure. This study was performed to evaluate the risk of urinary tract infection related to catheter insertion and the benefit of using prophylaxis antibiotics prior to catheter insertion. A retrospective study for 100 elderly patients from skilled Nursing facility and home care was done. The study reviewed the files and medical records of the patients between May 2011 to May 2012. The study examined incidence of UTI after catheterization and whether the use of prophylaxis antibiotic necessary before insertion of urinary catheter. The study revealed 72 patients out of the 100 patients enrolled in the study (72%) were free of UTI after insertion of urinary catheter. 28 patients out of 100 (28%) have suffered from UTI after the insertion of urinary catheter. 25 patient out of 28 patients who had UTI had the urinary catheter for more than 2 weeks. 25 out of 28 UTI patients have not received prophylaxis antibiotics, 3 out of the 28 patients have received antibiotic prophylaxis and suffered UTI. The authors concluded that Urinary catheters are commonly used among elderly patients. There are several reasons for the use of urinary catheter. The Most common is urinary incontinence, bedsore protection, urinary retention, and output monitoring. There is no need for using prophylactic ABX before or after the insertion of urinary catheter for the elderly patient. Insertion of urinary catheter under aseptic condition is important to prevent procedure induced urinary tract infection.

A review paper looked at inherited Ataxia's. The hereditary ataxias are a heterogeneous group of diseases. Most attempts at classification have been based on pathologic findings and are not always useful for the clinicians. Many of these disorders are multisystem degeneration in which the underlying biochemical or other defect is usually unknown. The pathophysiology is correspondingly poorly understood. Hereditary ataxia can be divided into the hereditary congenital ataxia, the ataxia linked with metabolic disorder, and early onset ataxia of unknown etiology (Table 1). Primary care physicians should be aware of the differential diagnosis of the hereditary ataxia when faced with ataxia of unknown etiology.

# Ageing and Security in Tehran, Iran:
# A Sociological Assessment

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## ABSTRACT

The paper investigates life security in the course of ageing in Tehran City which is generalizable to other parts of the country. The present survey studies various dimensions of the lives of the elderly people their material quality of life, their wellbeing, their abuse etc. The author tries to reflect on the quality of human rights and the dignity of the ageing people in the present paper. Though the number of elderly people is increasing in Iran, yet not enough institutions have been built to meet their increasing needs. The method of research used, is mainly empirical, yet it is preceded by theoretical and literature review. While five hundred elderly people have randomly been selected for the study, the main hypothesis of the study is: "increasing security of the elderly positively enhances their quality of life". The paper concludes that the new generation of the elderly with new needs and expectations are quite different from those of the previous generations and age cohorts.

Key words: Security. Elder abuse. Isolation. Quality of life. Wellbeing.

## Introduction

Population ageing is an obvious outcome of the process of demographic transition. While the developed countries of the world have already experienced the consequences of ageing, the developing countries are facing them now. The present paper reflects on some of the dimensions of the elderly people's vulnerability in Tehran, Iran. While the recent emphasis on studies pertaining to the elderly in the developing world is mainly attributed to demographic transition, the deteriorating conditions of the elderly are results of the fast-eroding traditional family system in the wake of rapid modernization, migration and urbanization (Irudaya, Sankara, & Mishra, 2005). This has in turn caused the elderly in almost every society to face challenges and various forms of insecurity. In Iran too, conditions, such as isolation, loneliness, physical weakness, uncertain future, etc., contribute to the sufferings of the elderly people as a whole.

Abuse of the elderly or elder abuse was publicly acknowledged in the 1980s. It takes many forms, from passive neglect to active torment, and includes verbal, emotional, financial and physical harm. As the proportion of elderly people rises, so does the incidence of abuse.

Most older people get support and comfort from their families, but some people face exploitation and abuse. No single term refers to abuse and neglect "any action or inaction by any person, which causes harm to the older or vulnerable person" (Council against Abuse of Older Persons, 2002).

Abuse against elder persons includes physical abuse, neglect (active or psychological passive), institutional abuse, and domestic violence. However, most abuse against elder persons comes from family members. Some theorists say that abuse against elder persons fits into a larger societal pattern of ageism and devaluation of elder people (Harbison, 1999).

As in other parts of the world, there has been an enormous growth in the elderly population in Iran also. Until the early 18th century, global population size was relatively static and the lives of the vast majority of people were "nasty and short".

Since then, the size and structure of the global population have undergone extraordinary change. More than three decades have been added to life expectancy, with a further gain of one or two more decades commonly projected for this century (Bloom, & Canning, 2008).

If we look at the global ageing scenario, we would find that the population of the world, which was 5.7 billion in 1995, is expected to reach 10.8 billion by 2050—a two-fold increase. In this fast growing population, the percentage of the elderly will also increase rapidly, i.e. from 9.5 in 1995 to 20.7 in 2050. Further, amongst the elderly, the number of the oldest ones—those aged 80 and above will increase more rapidly. That is, from 320 million in 1995 to 1,050 million in 2050 (UN World Population Projections, 1998).

So far as Iran is concerned, today, nearly 4,500,000 elderly people live in Iran. According to Iran's national census in 2006, about 7.27 percent of the total population of the country, and 6 percent of Tehran's population were over 65 years of age. With regard to the current demographic structure, it is anticipated to have an unprecedented increasing trend of population ageing in the years to come, i.e. from 7.27 percent in 2006 to more than 16 percent in 2050 — a phenomenon that has never happened before. Besides, since Tehran City has a good record of in-migration flow, the ratio of ageing population will be higher than the other urban areas in the coming years. It may also be pertinent to mention here that those born in the early 1940s; at the outset of Iran's modernization process, are at present increasingly reaching ages 65 and over, and are usually much concerned about their economic security, safety, marginal participation, and economic independence. This has also become a priority area for social welfare planning in Iran.

Table 1 below presents comparative trends of the elderly growth in Iran and Tehran City between 1956 and 2011:

As the Iranian society in general and Tehran City in particular are transforming, the characteristics of modern mega-cities are also appearing in them. Similarly, as the families are also transforming from traditional to modern, the living space is becoming very small and tight. In this process, the elderly face insecurity and other vulnerabilities. Besides, factors such as social,

psychological, economic and cultural changes also influence the different dimensions of the lives of the elderly, including their social security, welfare, safety, social cares/services, and ageism.

In this backdrop, the issues pertaining to ageing, elderly protection, residence quality, food conditions and their social networks are of prime importance. As a whole, the study intends to reflect an objective picture of the elderly people in Tehran.

In this sociological appraisal, apart from studying the physical, material and emotional well-being of the ageing population, their socio-cultural similarities and differences will also be appraised. The scenario, therefore, considers the elderly's quality of life from a variety of perspectives, such as demographic foundations, social and family relations, economics, health, disability etc. While the elderly with better economic conditions have more social security in their old age, the impoverished elderly largely face social insecurity. Therefore, paying attention to the economic and social security of the elderly, particularly the women, with regard to their health and treatment services, is of prime importance in both urban and rural areas (UNFPA, 1999).

The present study aims at identifying the quality and quantity of different age-groups, including those moving towards the old age, which would help the social planners to be familiar with this hidden phenomenon that has not much been touched by any governing body so far. It is expected to improve the quality of life of these people from the perspectives made in this study. For the purpose of this research, the 'elderly' age-groups of 60/65 years of age are included. Such people usually have common characteristics (Turner, 2000). The study would not only prove useful to students of gerontology, social sciences and welfare studies, but also help social workers, urban planners and others in understanding the quality of life of these people in a better way, their deficiencies, weaknesses and other dimensions of their vulnerabilities. It could also be a beginning for further studies in this field at national level.

In the present paper, consequences of change in family as a unit, and in the living style of the elderly, their expectations, and relevant sociological theories will be analyzed. Besides, social policies necessary for the elderly will be discussed at length.

**Table 1: Comparative Trends in Elderly Growth in Iran and Tehran City between 1956 and 2011**

| Year | Population | People 65 and above | % | Year | Population | People 65 and above | % |
|------|-----------|---------------------|-----|------|-----------|---------------------|-----|
| 1956 | 18954407 | 758670 | 4.0 | 1956 | 1560434 | 52013 | 3.3 |
| 1966 | 25788722 | 993045 | 3.8 | 1966 | 2719730 | 81295 | 3.0 |
| 1976 | 33708744 | 1186470 | 3.5 | 1976 | 4536264 | 14829 | 3.3 |
| 1986 | 49445010 | 1493382 | 3.0 | 1986 | 6010075 | 203062 | 3.4 |
| 1996 | 60055488 | 2587437 | 4.3 | 1996 | 6758845 | 320430 | 4.6 |
| 2006 | 70472846 | 5130000 | 7.3 | 2006 | 7803883 | 464638 | 6.0 |
| 2011 | 75149669 | 434304 | 5.8 | 2011 | 8154051 | 604283 | 7.5 |

**Source: Results of the Population and Housing Censuses of Iran, 1956-2011.**

Though policy makers and social researchers themselves, too, are prone to sufferings arising from old age, in this regard, they do not often project their own future and loneliness (Shamloo, 1985).

## Method of Research

The focus of the research is to present a combination of theoretic frameworks and empirical realties. For the empirical part of the study, 500 elderly people were randomly selected from different parts and neighborhoods of Tehran City. They were approached through direct interviews with the help of pre-designed questionnaires. Eventually, the filled-in questionnaires were edited, electronically extracted and presented in the form of tables. The researcher also reviewed the background literature on ageing population from different societies of the world. In the theoretical part, the researcher tried to refer to, and make use of sociological and gerontological theories within reach.
The main hypothesis in this research is *"increasing security of the elderly positively enhances their quality of life"*.

## Findings

Among the respondents, i.e., 500 elderly people, who were referred to, and surveyed in various parts of Tehran City on a random basis, 89 (17.8%) were in the age-group 60-62, and 99 (19.8%) were in the age group 63-65. Gradually, and with increase in age, the ratio and number of ageing people finds a declining trend:  in such a way, only six (0.5%) of people within the samples were found in the age-group 90 and above. The age structure further shows that the sex ratio for the elderly (i.e., 60 years and above) is 106 . This high sex ratio for the elderly is due to high fertility and high maternal mortality for the women in the past.

The survey also found that the elderly people with spouse were 60 percent, whereas the percentage of elderly widows was 29, male elderly 8.4 percent, and finally those never married were 2.6 percent of the total elderly studied in various neighborhoods of Tehran. Similarly, 55.8 percent of the respondents enunciated that they never received any financial assistance from their sons, whereas 19.2 percent received a little bit. Likewise, 18 percent declared that they were reliant on their sons' assistance, and finally 7 percent stated that they received a considerable amount of financial assistance from their sons every month. In Iranian culture, parents normally expect to be financially assisted by their sons in their old age. The above figures indicate that most of the elderly are facing financial constraints despite having male offspring. So far as the emotional attachment of the sons is concerned, almost 20 percent of the respondents said that they were highly attended by their sons, whereas 44.6 percent said that they were much benefited by the emotional support of their sons, and 35.2 percent declared that they were not absolutely attended by their sons. The figure indicates that sons did not care for their elder parents at all. However, government support and help by social workers can to some extent reciprocate this lack. That is to say, due to such failure, social workers provide the elderly with the necessary assistance.

Results of the survey further show that financial support by daughters towards old parents is very insignificant. While a little less than two-thirds, i.e., 63.4 percent, of the respond-

ents declared that they did not receive any financial help from their daughters, 20.4 percent asserted that they got a little bit, and only 16.2 percent were satisfied with such help from their daughters. Such a scenario strengthens the idea of son preference in the Iranian society, and this heritage passes to the future generations too. However, daughters emotionally attend to their old parents very much, as less than three-fourths, i.e., 73 percent of respondents gave an affirmative answer in this regard, whereas only 27 percent replied in negative.

Generally speaking, such dissatisfactory help and attention from offspring to parents, particularly in the old age, and towards people who are highly traditional in their lifestyles and attitudes, creates problems more due to the emerging generation gap. Although literacy can help the elderly to get themselves engaged in reading books and newspapers, something like 27 percent of the respondents were not literate at all. This percentage was 16.7 for the male elderly and 37.8 for the female elderly. However, in the present study, about 20 percent of the male respondents had higher qualifications, while this ratio stood at only 1.6 percent for the female elderly.

The elderly people usually have plenty of leisure time, and that is also reflected in the present study. While about 48.2 percent of the respondents preferred to watch television during their free time, 29 percent read books and newspapers, and the remaining 23 percent used other means of entertainment. Similarly, while the rate of reading newspapers by male samples as leisure is 22 percent, 55 percent of female samples assert that they watch television as their leisure.

The results of the research show that little less than two-thirds of the elderly, i.e., 64.6 percent,  had mostly interrelations with their children, whereas 14.5 percent had it with their friends, 9.4 percent with relatives, and finally 11.5 percent with neighbors and others. While one of the psycho-spiritual needs of the elderly people is known to be travel and tour, more than 20 percent of them had not traveled at all during the period of study. It is worth noting that the elderly people in the Western countries usually travel very frequently.

Within the samples of the present study, the elderly were usually found satisfied with their access to medical services. To a question about their health and safety, only 39.3 percent of them declared that they were safe and healthy. Health demography is very much associated with safety and security. On the other hand, 25 percent informed that they had arthritis problems, 10 percent of them had diabetes, and 27.4 percent had other ailments. In the course of inquiries, it was also found that 91 percent of the respondents were not interested at all in staying in nursing homes.

## Discussion

The basic concept of social security is not new to Iran. Traditionally, a sort of traditional support system (e.g., joint family system) existed before, to provide security to elder destitutes and other vulnerable groups in society. In such circumstances, family was the protector of the elderly, and thereby, the elderly did not have to resort to the secondary institutions, such as nursing homes, for their protection. However, such a system is

gradually disappearing, and a state-based social security system has come into being, but it is not adequate for a large chunk of elderly population. Findings indicate that the offspring still feel responsible to help their elder parents as much as they can. For example, 18 percent of the respondents receive monthly financial help from their sons, and 7 percent assert to be fully assisted by their sons per month.

Iran, which is comprised of 31 provinces and 1148 towns and cities (SCI, Tabnak News, 2015), does not have the conditions of a balanced development; in that, some provinces are more developed, than others. Under such conditions, some of the elderly people living in affluent provinces are in improved and better conditions, whereas others in deprived provinces need to be more attended and cared for. In the latter provinces, they are in a state of more insecurity and vulnerability. For example, Tehran Province, especially Tehran City, is reckoned as a more secure site for the elderly people. In Tehran, the elder people have more facilities with regard to public and private nursing homes, easier access to medical facilities etc. They have more and easier access to parks, hospitals and other sites which can provide them with the required services. Therefore, under such circumstances, men and women in other parts of the country tend to migrate to this province, particularly Tehran City, to be more benefited of care and security in their old age. Similarly, as larger cities attract more social and material capital in Iran, they are more developed, and hence are provided with more facilities. Such perspectives attract more elderly people; often accompanied by their children.

Because of young population structure in Iran, almost 60.5 percent of the entire population of the country stand at age groups 0-29 (Iran Population Census, 2011). This young population is prioritized in different sectors of planning and housing investments. Thus, smaller investments on housing and health sectors are done in favor of the elderly. For instance, while a great number of the elderly people in various parts of the country are in immediate need of protection centers, they cannot easily have access to such facilities. They have to either live alone, or be a burden on their offspring. Nevertheless, it is hoped that more priorities would be accorded to the elderly community in the sixth five-year plan scheduled to start in 2017.

Based on inferences and ad hoc interviews with a number of elderly people, they do face many problems as far as their protection and care are concerned. However, the elderly with poor socio- economic backgrounds face multiple problems and constraints. What is therefore required as an immediate step towards an improvement in the elderly's quality of life, is the establishment of centers for the elderly in all regions, which could meet their various needs, such as habitation, care and food. In the meantime, the negative attitudes of the people towards nursing homes must be changed in an acceptable manner, so that they could access improved social and economic lives.

I must add that transformation of the economic structure of Tehran City in the past few decades—from agricultural to industrial and service sectors; in the wake of urbanization, has, to a great extent, changed the attitudes of families towards the elderly members, and the quality of protection that they provide. They have now largely recognized nursing homes as a social fact and

as a positive alternative to keep and protect the elderly. Younger family members being involved in modern civil life, they have implicitly come to recognize nursing homes as positive places to accommodate, and attend the elderly. Increase in life expectancy rate in 2015 up to 72 years for males and 76 years for females in Iran (World Population Data Sheet, 2015), represents a good outlook of ageing in the country, which requires appropriate planning in both rural and urban areas. Based on table 1 in the present paper, life expectancy has highly contributed to increase in percentage of the elderly people between 1956 and 2006. Yet, increase in longevity is unjustly distributed among various social strata in Iran. However, such a transition contributes to rising needs of housing, pension, medical care, etc., within the growing elderly citizens. Generally speaking, this scenario has contributed to the Iranian elderly people gradually facing the loss of inter-generational solidarity and integrity. Somehow or other, results of the study confirm the emergence of the scenario.

Based on the research conducted in different parts of Iran including Tehran City, there are considerable differences in the quality of life within elderly males and females. Males are found to be in a comparatively better position. The differences stem mainly from more wealth that they hold, possibility of re-marriage after losing spouse for males, more social respect for the male elderly etc. Such opportunities more await male ageing people rather than the females.

## Theoretical Concepts

In the past, not only in Iran, but in many other societies also, the elderly people were considered as sources of knowledge, rationality and experience. Currently, with the passage of time, and as a consequence of the social, economic and cultural changes taking place in all societies, they have gradually lost their importance and charismatic values. While in the 19th century, and even in the early 20th century, grandchildren rarely had the chance of seeing their grandparents, now and particularly at the outset of the 21st century, with the rise in average age, three or more generations have got the opportunity to simultaneously live together.

Physical changes that spring up in people in the course of time, are not necessarily displayed among all people of the same age-groups in a similar manner, but their physical and mental effectiveness, and their reactions towards such factors, are different within different age-groups of elderly people. In fact, the elderly suffer one (in some cases, three at the same time) treatable sickness(es) (Keddie, 1998).

From the demographic point of view too, the number of the ageing people in Iran is increasing, and the scenario needs the provision of more medical care, sufficient nutritious foods, and other necessities of life. Under such conditions, special social policies should be made and applied for these elderly people, and any failures in this regard would lead to problems for them, and consequently they would reflect as a burden. In other words, though entering old age has been identified as ages 60-65, i.e., the age in which people usually retire, yet, we cannot apply a definite age border in which the people should be named 'aged' (Akbarzadeh, 1997). In another definition, old age has been called repeated dependency, i.e., a period followed by the period of dependency and socialization, followed by the second period,

i.e., independence and maturity, and eventually the 'Third Age' or the beginning of the elderly period (Laslett, 1998).

Census data indicate the sex ratio of 108 for those elderly people aged 80 and over (Population Census of Iran, 2011). Therefore, with regard to increasing female elderly, more arrangements should be made to protect them in their old age. Elderly women being poorer than men due to their limited sources of income in their old age, need to be more protected and attended.

In a global comparison, in the developed world, while 17 percent of the people are aged 65 and above, the ratio stands at only 6 percent in the developing world (World Population Data Sheet, 2015). In this regard, Iran's ratio of ageing population stands at 7.3 percent, and that of Tehran City stands at 6 percent (Iran Population Census:2006). On the other hand, while because of ageing, there happens a sharp fall in people's safety, and people's social costs, their medical and health costs increase (IFA, 2015). In this regard, creation of supportive establishments at neighborhood levels would be very effective and enabling.

## Social Isolation of the Elderly People

One of the main dimensions of social insecurity and vulnerability of the elderly people stems from their social isolation. In fact, isolation is one of the general issues of old age, and that is the result of the elderly's non-functionality or lack of role in the society. However, it is extremely difficult to measure or define social isolation (Shanas, 1968). It may be objective (e.g., social contacts can be counted), or it can be subjective (e.g., people can be asked about their feelings). Most studies on social isolation have been concerned primarily with the four types of isolation as outlined below:

• By comparison with their contemporaries in the developed world: peer-contrasted isolation.
• By comparison with younger people: generation-contrasted isolation.
• By comparison with the social relationships and activities enjoyed by (other younger or middle-aged people): age-related isolation.
• By comparison with the preceding generation of old people: preceding cohort isolation.

Measurement of isolation usually consists of information about social activities as a way of estimating the number of social contacts (Townsend, 1968).

Quite a lot (one-fifth) of those interviewed in the present study were found isolated, or extremely isolated, and the majority of them included women (widowed). This high percentage of isolation within the elderly people is more due to the breakdown of joint family system, weakening of family network, and more migrations by the younger family members in Iran in current times as a whole. Isolation does not necessarily increase with age, but there is little variation in the likelihood of being visited by family, friends and relatives. The appearance of this phenomenon is highly due to the complexity of life in highly populated Tehran City with a poor and congested transportation network. Though the two concepts of isolation and loneliness are almost synonymous and are used interchangeably, yet, there is a conceptual distinction between the two. Isolation relates to circumstances (which can usually be measured, however crudely), whereas loneliness relates to feelings (often about these circumstances). Widows are also likely to experience loneliness as do others who have been bereaved (Bury & Holme, 1991).

The problems of isolation and loneliness not only arise within the elderly people who are dispersed in a city, but they may also happen to the elder people residing in nursing homes. Studying the older people in residential care, Townsend (1986), has commented that "the lack of even a single friend, a higher rate of severe or frequent loneliness, the discouragement of spontaneous social activity, the inability of visiting relatives and friends to adopt useful roles, the lack of satisfying, and sociable occupation, all represent isolation and loneliness by the elderly even in care centers."

Social isolation is more problematic for women, since they usually live longer than their male counterparts. Culturally speaking, women usually have less outside-home relationships and contacts in Tehran City as compared with men, and that contributes to more likelihood of isolation among them. Similarly, as mentioned earlier, women in Tehran City have longer life expectancy than men in this city. Such a phenomenon makes them highly vulnerable in various grounds and spheres. Under such conditions, women are more prone to psychological sufferings like dementia. While sociologists highly recommend that the elderly people should have their own independent lives, yet providing them with adequate financial resources remains unsolved in many instances. Clinicians have identified a broad array of risk factors typically associated with elder abuse and neglect (Podnieks, 2004).

Factors such as caregiver's mental health, substance abuse, dependence on the care recipient etc., have been described as important indicators leading to social isolation of the elderly people. Similarly, widowhood, the deaths of friends or children moving away can lead to social isolation (Hall & Havens, 2002). Such circumstances widely relate to Tehran City too.

To get rid of the problem of isolation among the elder people, social networks can prove to be very important. These networks may include both the fellow co-residents as well as from beyond the household, including friends, neighbors, etc. So far as the elderly in Tehran City are concerned, there are still further steps to be taken in their favor. For example friendship centers are to be established in neighborhoods to help them with isolation.

## The Elderly's Housing

Adequate and suitable housing is also reckoned as an example of the elderly security, and the provision of which would bring safety and security to the weak and disabled elderly people. However, there is a growing recognition of the link between poor health and housing. Features such as the absence of stairs/ramps may force even a very frail person to continue living independently. Many old people, however, live in less than desirable housing and some of them have particular problems. There is evidence too, that some people remain in residential care only because alternative accommodation is lacking (Wagner, 1988).

In the course of interviews with the senior respondents in Tehran City, the researcher came to know that almost 60 percent of the interviewees preferred/wanted neighbors without children. The study found that such housing sites give elder people more feelings of well-being - giving them more control over their social contacts.

Though the problem of elderly's adequate and suitable housing has to some extent been tackled in the Western societies, it still does exist in the developing nations, including Iran. If we look at the basic family structure in Tehran City, many parents are found to live with their children, as joint families. Therefore, the elderly's housing quality tends to depend on the economic capacity and shelter patterns of the household. It is worth noting that the trend in population ageing (growth) in Iran is even faster than its socio-economic growth rate. Thus, if necessary arrangements are not made for the elderly in a futuristic manner, the gap/difference will contribute to further challenges for the elderly. In view of this, dealing with this problem in urban areas has been a priority for the large City of Tehran.

So far as preferences in apartment living is concerned, no single type of housing fits all older people in Tehran. But, older people do prefer some types of apartments and neighborhoods over others; depending on their middle-age lives and backgrounds.

## Types of Insecurity

Although there has been a rise in the all-age mortality from accidents, the number of deaths from accidents in those aged 65 and above is fairly constant. In the course of study, the researcher came to know that the elderly people are frequently at risk of different conditions.

Older people, like people of other age-groups, suffer domestic accidents, fires, burns, poisoning, road/workplace accidents and falls.

• Older people are at particular risk from injury or death from domestic fires.
• Falls account for almost one-third of deaths from all accidents. Older people are at particular risk with 65 percent of fatal falls occurring among those aged above 75.

## Risks Factors and Recommended Strategies

The risks factors that the researcher acknowledged are associated with accidents and injuries among old people are very broad, and include the following:

• Environmental factors
• Inappropriate use of medicine
• Poor health status
• Disability
• Being overweight
• Lack of exercise
• Nutritional deficiency
• Poor housing
• Lack of training for careers

However, as the number and proportion of the elderly people in Iran is growing; especially those living alone, they may be more vulnerable to accidents. Therefore, greater attention should be paid to their safety and security both inside and outside home. Thus, a number of strategies aimed at these people should be implemented:

• Extended awareness programs for elder people, their careers and health professionals, associated with accidents.
• Promotion of healthy ageing practices, such as reduced smoking, increased exercise etc.
• Educational programs to improve knowledge and safety skills.

## Conclusion

In this research, a multidimensional perspective of the elderly people's lives in the capital City of Tehran, as part of the macro society of Iran, is presented. The research reflects that the 'new elderly' (those born in the early 1950s) have even more concerns as compared with the 'older elderly', or a generation before them. Tehran City, being a migrant-receiving city, while the ratio of its in- migrants is higher than other cities, the expectations of the elderly people in this city are by far different from those in other cities. Under the conditions of modern life, elderly's general welfare, care/protection and financial constraints are quite remarkable in different neighborhoods with special reference to the current socio-economic conditions.

Tehran City is considered as a part of the country wherein the elderly are more prosperous. They have easier access to medical care, nursing homes etc. Based on research done in various age-groups, marked variations have been found with regard to the quality of life of male and female elderly people; stemming from more wealth within the reach of males, possibility of remarriage by the male elderly and so on. While the improved socio-economic conditions have made it possible for three generations to live together, yet, modern life has also forced them to live away from their families. Regardless of class, race, creed and color among the elderly, what is common among them, is disability and dementia.

While this research has considered and analyzed some of the challenges, profiles and problems of the elderly people, yet, the scenario of ageing needs more research studies on a regular basis in the future too.

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# Antibiotic Prophylaxis to prevent Urinary Tract Infection following Urinary Catheterization in Geriatrics

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## ABSTRACT

**Background:** Urinary Catheterization is a common procedure in the elderly population. Literature showed that up to 40% of the nosocomial infections are caused by urinary tract infection related to urinary catheterization. Some studies suggest using prophylactic antibiotics prior to insertion of urinary catheter in the elderly to minimize the chances of urinary tract infection related to the procedure. This study was performed to evaluate the risk of urinary tract infection related to catheter insertion and the benefit of using prophylactic antibiotics prior to catheter insertion.

**Methods:** A retrospective study for 100 elderly patients from skilled Nursing facility and home care was done. The study reviewed the files and medical records of the patients between May 2011 to May 2012. The study examined incidence of UTI after catheterization and whether the use of prophylactic antibiotics was necessary before insertion of urinary catheter.

**Results:** The study revealed 72 patients out of the 100 patients enrolled in the study (72%) were free of UTI after insertion of urinary catheter. 28 patients out of 100 (28%) suffered from UTI after the insertion of urinary catheter.

25 patients out of 28 patients who had UTI had the urinary catheter for more than 2 weeks.

25 out of 28 UTI patients did not receive prophylaxis antibiotics, 3 out of the 28 patients received antibiotic prophylaxis and suffered UTI.

**Conclusion:** Urinary catheters are commonly used among elderly patients. There are several reasons for the use of urinary catheter. The most common is urinary incontinence, bedsore protection, urinary retention, and output monitoring.

There is no need for using prophylactic ABX before or after the insertion of urinary catheter for the elderly patient.

Insertion of urinary catheter under aseptic conditions is important to prevent procedure induced urinary tract infection.

**Key words:** Antibiotic prophylaxis, UTI, Catheterisation, geriatrics

## Definition

Urinary tract infection is the presence of bacteria in urine, kidney, urinary bladder, urethra or prostate.

UTI is one of the most common infections in the elderly.

Urinary catheter (tube inserted through the urethra to drain urine from the bladder to urine collecting bag).

Urinary tract infection may occur when the elderly patient catheterized for a prolonged period (>2 weeks). The colonized organisms may invade the mucosa of the urinary bladder and cause bacteriuria, cystitis and may cause bacteremia.

## Types of urinary catheters

**1.** Indwelling catheters (urethral or suprapubic) can be used for short and long term catheterization.

**2.** External catheters e.g. (condom catheters) used for men with dementia and pressure sores and have lower risk of infection.

**3.** Intermittent catheterization is used for short periods post surgically and in rehabilitation after spinal cord injury, and will be removed after emptying the urinary bladder.

## Causes of UTI during urinary catheterization

Urinary tract infection is a common disease in elderly patients. The prevalence of UTI approaches 40 % of nosocomial infections. According to the landmark studies 10-27% of the catheterized patients will develop bacteriuria within 5 days of hospitalization, 4% of the catheter related UTI will develop bacteremia. Most patients with long term urinary catheters are elderly.

There are many causes for contracting UTI in elderly, especially diabetics and immune compromised patients; among these are contamination and colonization of bacteria following urinary catheter insertion, residual urine in the collecting urine bags, bacteria from the bowel and backward flow from the catheter to urinary bladder.

UTI can be life threatening and cause severe complications in elderly. These can be prevented or reduced if proper hygiene is followed during and after insertion of urinary catheters and if the symptoms are discovered and treated early.

The most common bacteria that causes UTI in elderly are E-coli, Pseudomonas, candida albicans staphylococcus and enterococcus species.

Proteus and pseudomonas are the common causes of biofilm growth on the urinary catheters.

Many complications can occur as a result of urinary catheterization; among these are urinary tract infections, obstruction of the catheter, urinary calculi, local trauma, bladder fibrosis, prostatitis, pyelonephritis, local infection e.g. urethritis, local abscesses, tubule-interstitial nephritis.

## Background

This is a retrospective study of the need for antibiotic prophylaxis for preventing UTI following urinary catheter insertion in the home care services and skilled nursing facility from 1st of May 2011 till 1st of May 2012.

## The goal of the study

To evaluate the need of using prophylactic antibiotics before or after the insertion of urinary catheters.

## Epidemiology

**Place:** Home Care, Skilled Nursing Facility, Doha, Qatar.

**Duration:** 12 months.

**Date:** 1st of May 2011 - 1st May 2012.

**Patients number:** 100

**Method of data collection:** patients' files.

**Method of diagnosis:** The criteria that has been used for the diagnosis of UTI depended on symptoms like fever, abdominal pain, decreased urine output, change of urine color, hematuria and lab works like CBC, Urine routine and culture.

**Figure 1. Total number of the patients is 100 out of which 55 males and 45 females**
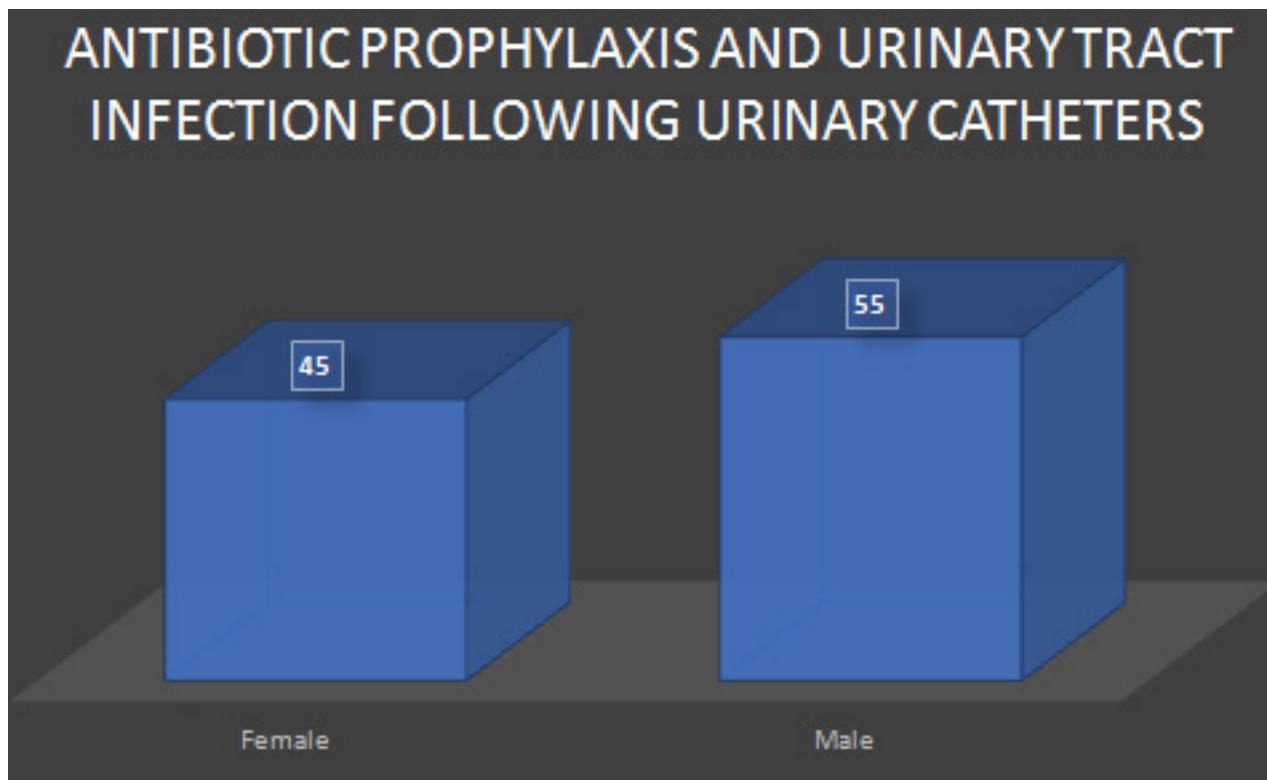


**Figure 2. shows the various reasons for urinary catheterization.**
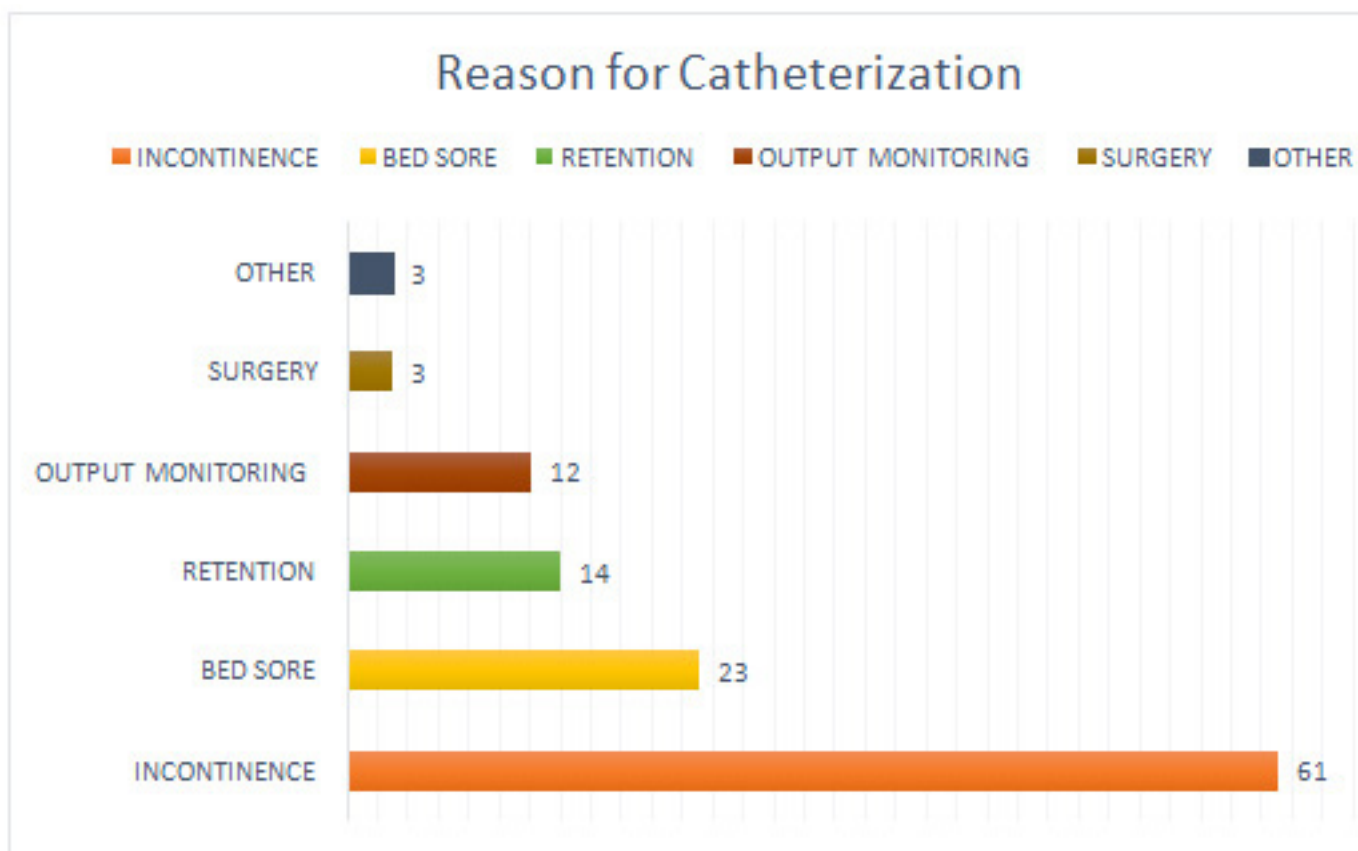


The most common reason for catheterization is incontinence and bed sores protection.

**Figure 3. shows the duration of urinary catheterization.**



DURATION OF CATHETERIZATION
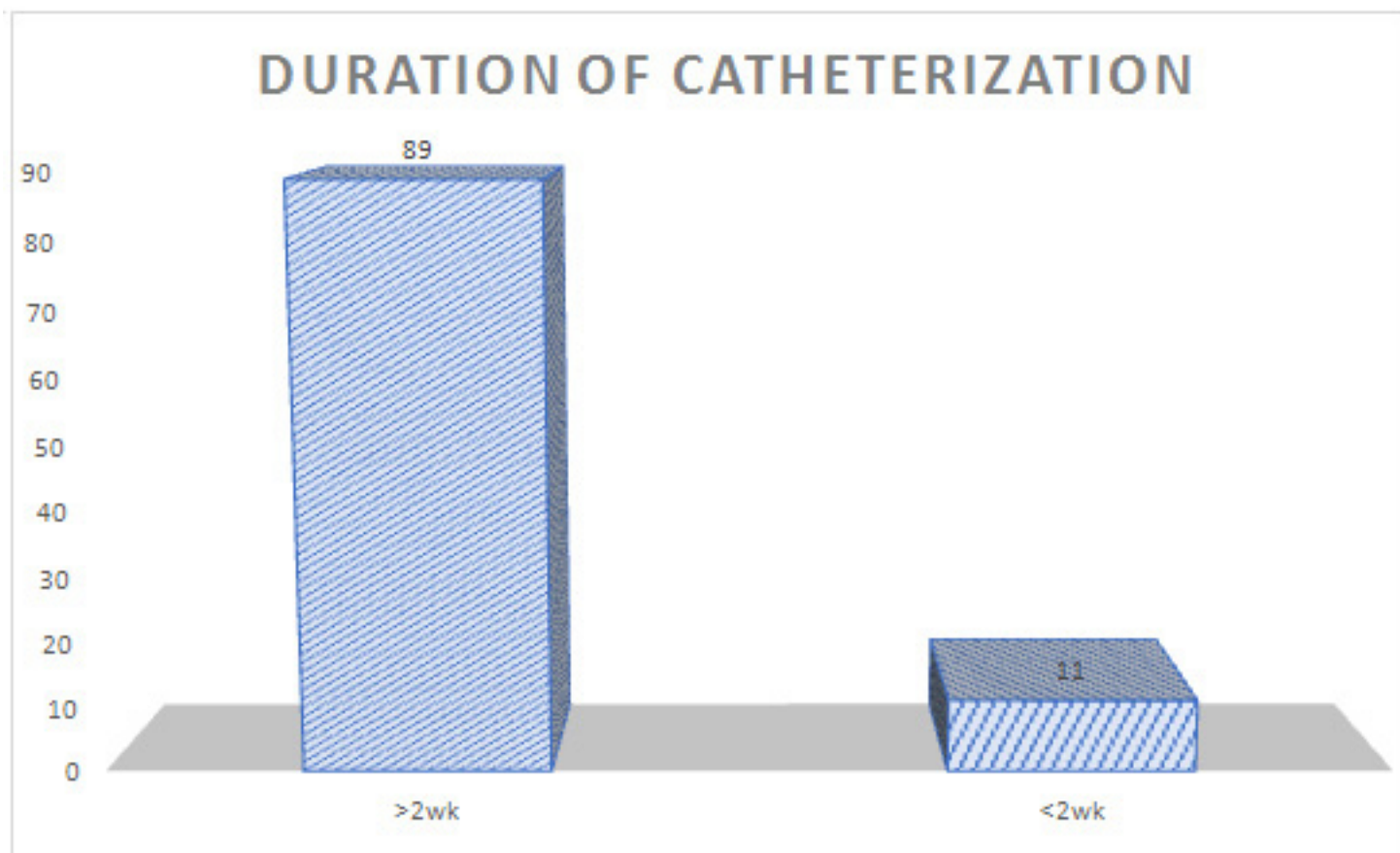
Most of the patients were on long term catheterization for more than 2 weeks.

**Figure 4. shows the number of patients who used prophylactic antibiotics.**



prophylaxis antibiotic used

**Figure 5. Illustrates the number of patients who suffered UTI after insertion of catheterization in the study.**



Most of the patients who suffered UTI were catheterized for a prolonged period (more than 2 weeks) which is also illustrated in Figures 7 and 9.

**Table 1. Illustrates male and female elderly patients who developed UTI after insertion of urinary catheter.**

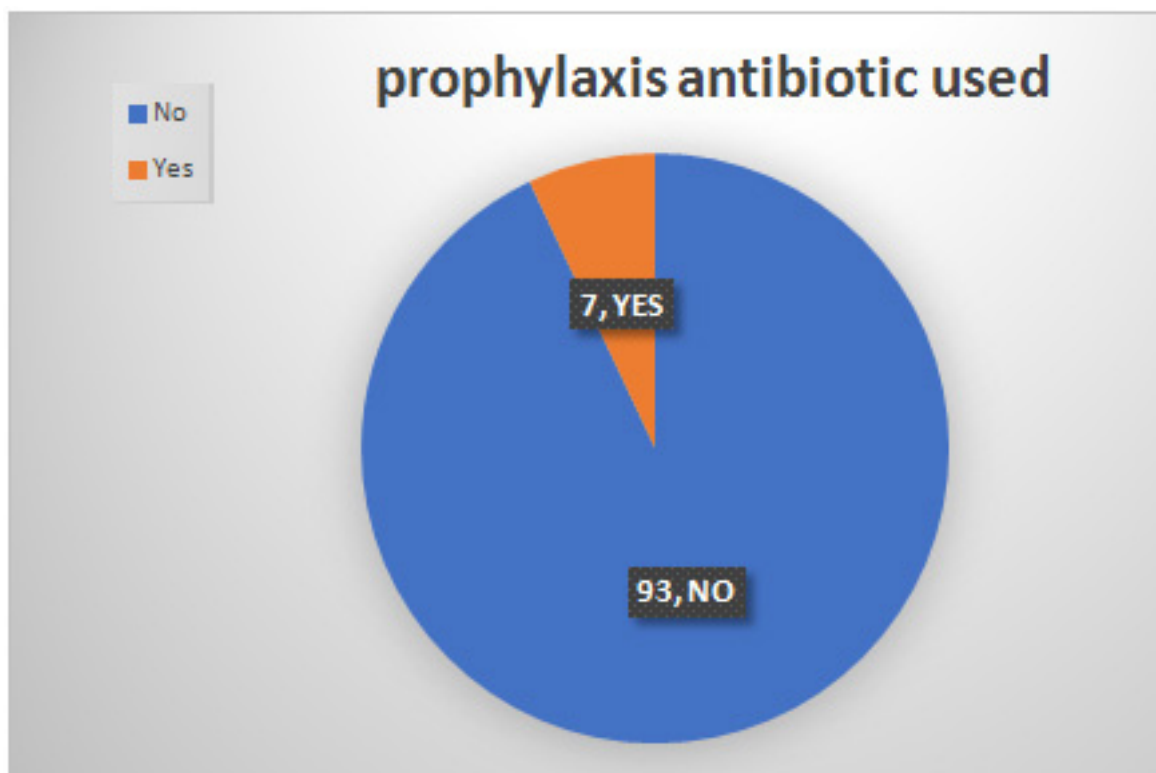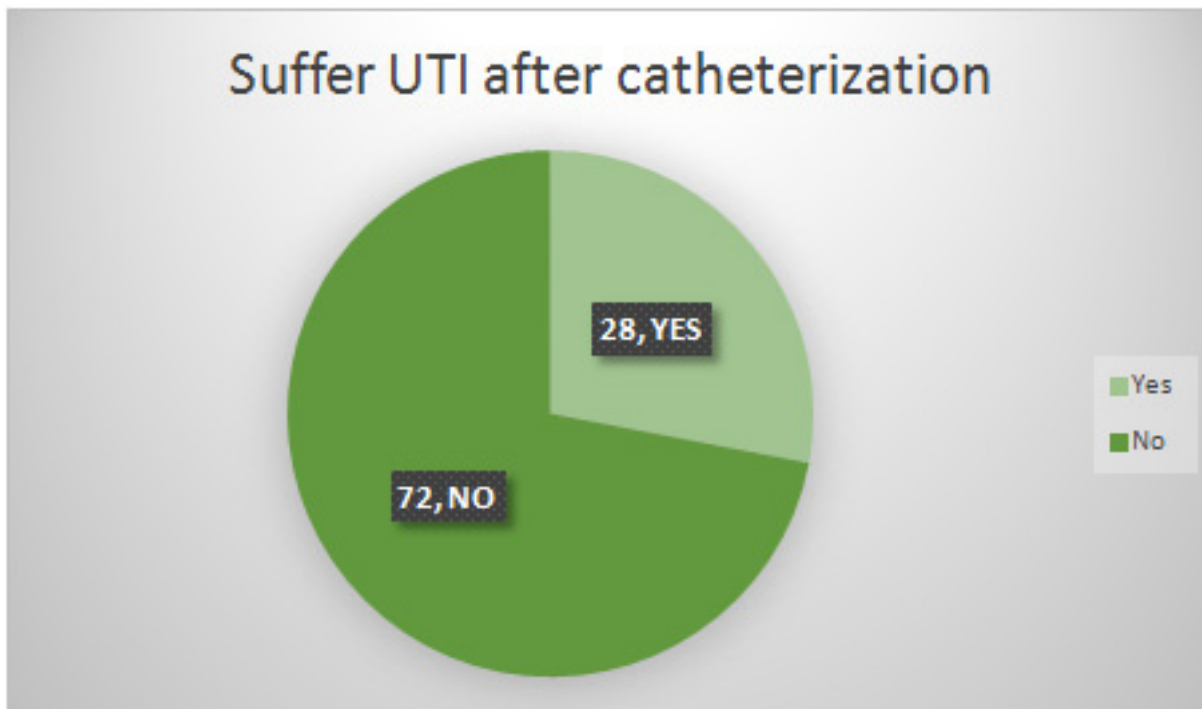| Number | Male | Female | Prophylaxix | > 2 weeks | < 2 weeks | Organism |
|--------|------|--------|-------------|-----------|-----------|----------|
| 1 | yes | | yes | yes | | E Coli |
| 2 | | yes | yes | yes | | E Coli |
| 3 | yes | | yes | yes | | E Coli |
| 4 | yes | | No | | yes | Pseudomonas |
| 5 | | yes | No | | yes | E Coli |
| 6 | | yes | No | | yes | Mixed growth |
| 7 | | yes | No | yes | | E Coli |
| 8 | yes | | No | yes | | E Coli |
| 9 | | yes | No | yes | | E Coli |
| 10 | | yes | No | yes | | Citrobacter F |
| 11 | yes | | No | yes | | Pseudomonas |
| 12 | yes | | No | yes | | Staph+Pseudomona |
| 13 | | yes | No | yes | | Proteus+Ecoli |
| 14 | yes | | No | yes | | Candida |
| 15 | | yes | No | yes | | Mixed growth |
| 16 | | yes | No | yes | | Pseudomonas |
| 17 | yes | | No | yes | | Pseudomonas |
| 18 | yes | | No | yes | | E Coli |
| 19 | yes | | No | yes | | Candida |
| 20 | yes | | No | yes | | E Coli |
| 21 | yes | | No | yes | | Mixed growth |
| 22 | yes | | No | yes | | Klebsiella |
| 23 | | yes | No | yes | | Enterobacter |
| 24 | | yes | No | yes | | E Coli |
| 25 | | yes | No | yes | | Candida |
| 26 | yes | | No | yes | | E Coli |
| 27 | yes | | No | yes | | E Coli |
| 28 | | yes | No | yes | | Proteus+Ecoli |

It also showed the duration of catheterization and type of organism found in urine culture.

**Figure 6. Illustrates the UTI causative organisms in male patients.**
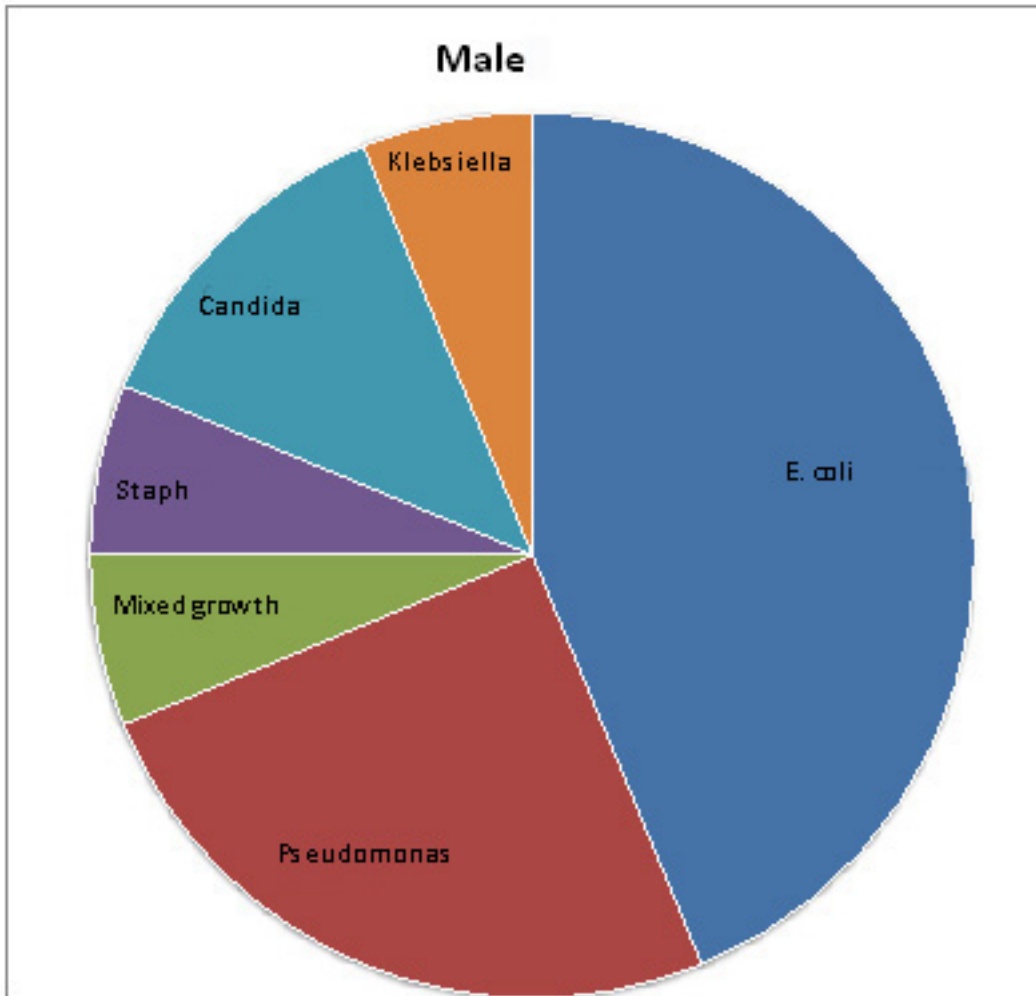


Figures 6 and 8 show the causative organisms of UTI in catheterized patients.

It shows E-Coli as the main cause in both males and females.

3 patients out of 28 have developed UTI even after receiving antibiotics prophylaxis.

**Figure 7. Shows the duration of urinary catheterization in male patients who developed UTI.**
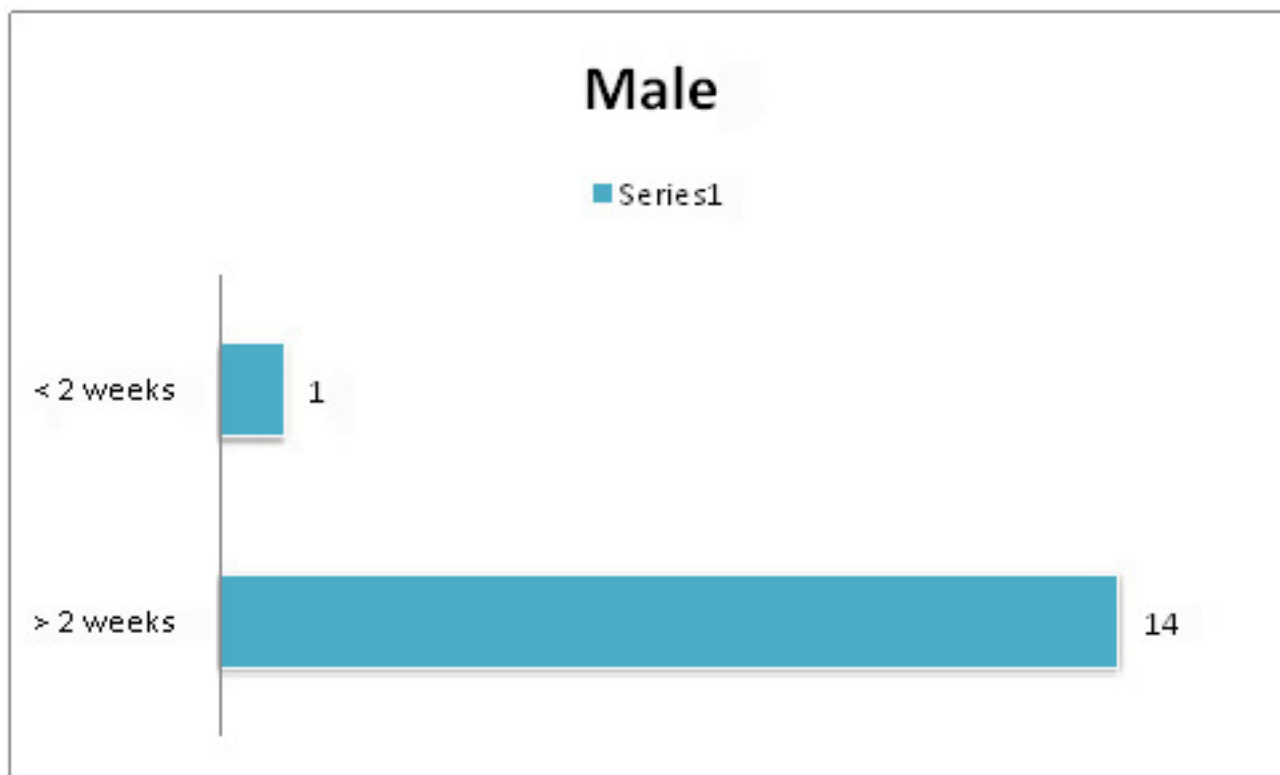


**Figure 8. Shows the causative organisms in female patients who developed UTI.**
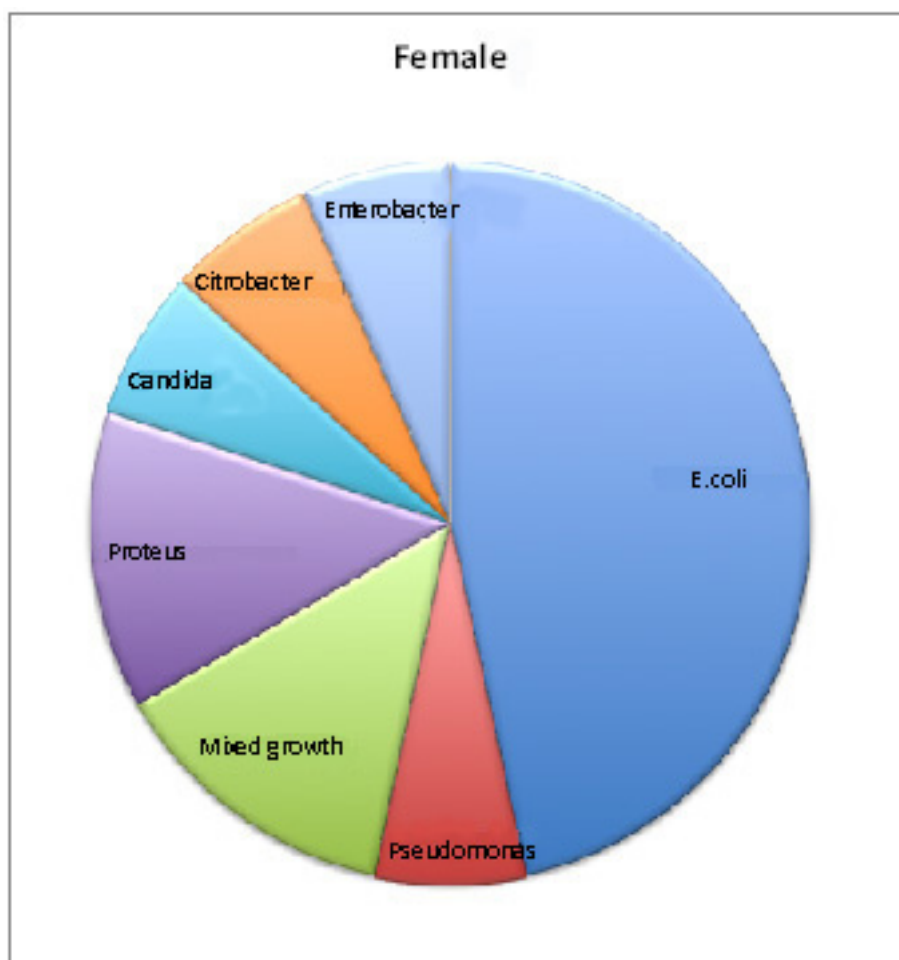
**Figure 9. Shows the duration of urinary catheterization in female patients with UTI.**
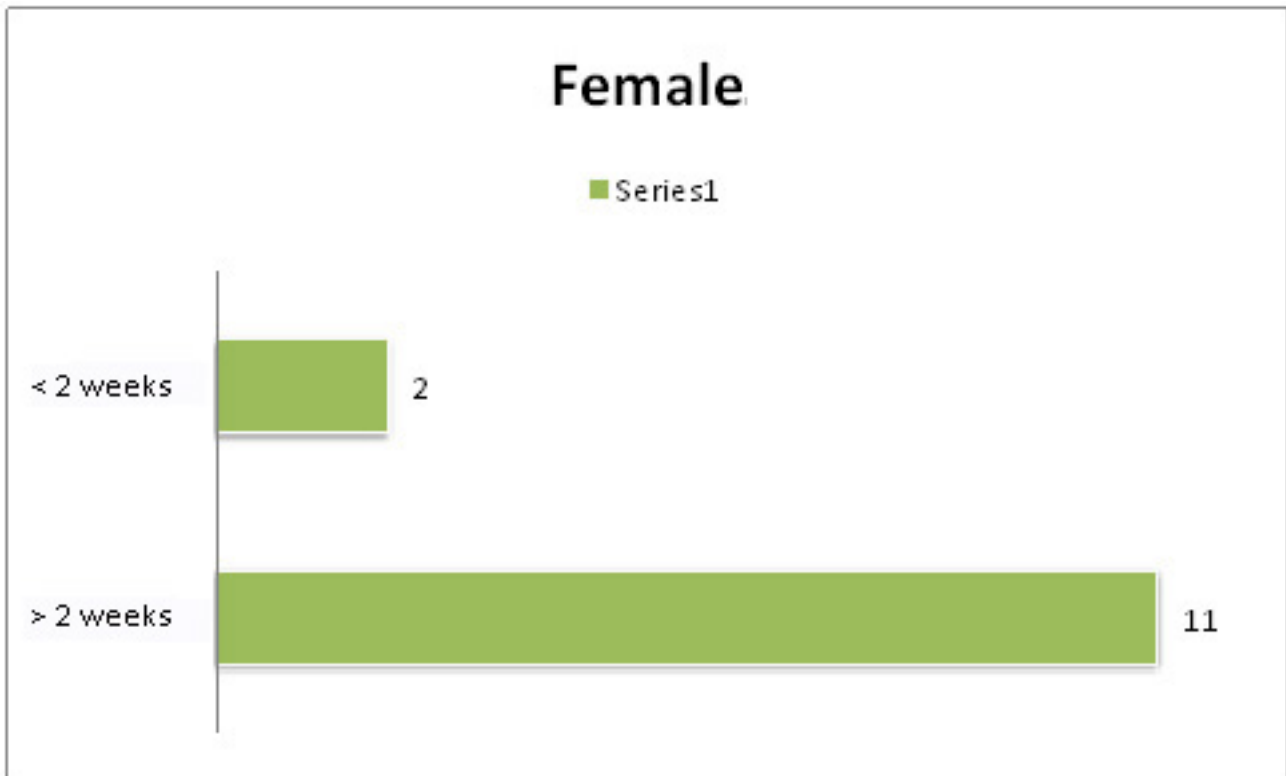


**Figure 10. Shows number of patients who developed UTI with or without antibiotics prophylaxis.**
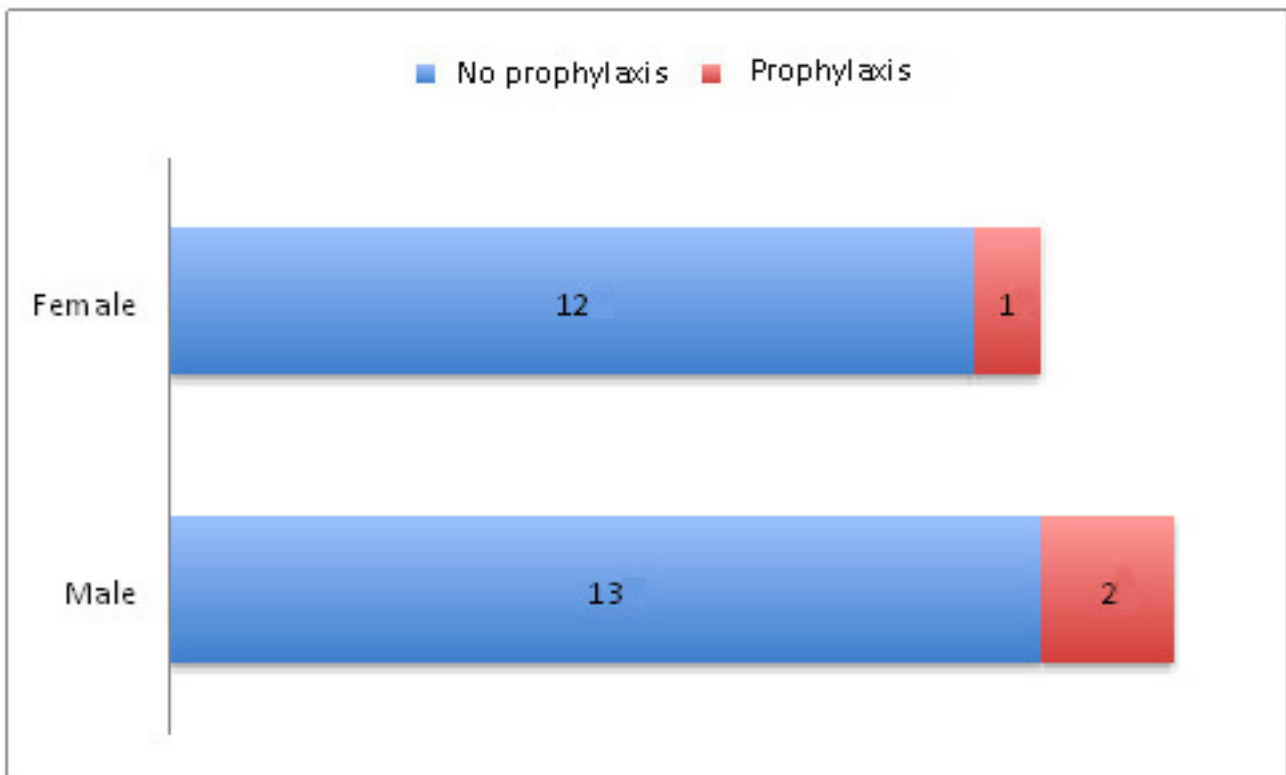
## Summary

Urinary catheterization is a common procedure in the elderly population especially in long term facilities.

Urinary tract infection is a common disease in the elderly.

The catheterized patients are prone to develop UTI especially with long term catheterization.
e.g. prolonged catheterization more than 2 weeks.

Prophylactic antibiotic use is not necessary to prevent UTI in case of catheter insertion in elderly.

Bacterial colonization is common in catheterized patients and this can lead to bacteriuria UTI and bacteremia.

UTI can be prevented if aseptic technique is used while inserting urinary catheter then followed with proper hygiene.

The most common organisms infecting urinary tract in catheterized patients are E-Coli, pseudomonas and candida albicans.

## Strategies to decrease UTI during catheterization

**1.** Aseptic technique during insertion and removal.

**2.** Daily catheter care e.g. cleans the skin around the tube.

**3.** Short term catheterization.

**4.** Remove the urinary catheter if no clear indication.

**5.** Proper regular urine bag emptying.

**6.** Prevent the urinary catheter kinking.

**7.** Keep the urine collecting bag below the level of the bladder.

**8.** Proper hand hygiene.

**9.** Replace the urine catheter regularly every month.

*Original Contribution/Clinical Investigation*

## Late onset Systemic Lupus Erythematosus: Tunisian cohort

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## ABSTRACT

**Introduction:** Systemic lupus erythematosus (SLE) is usually described as a disease that most often strikes reproductive-age women. However, SLE can be observed in children and in the elderly. Late onset of systemic lupus erythematous(SLE) seems to differ from young onset disease. We discuss the clinical course, immunological features, treatment and prognosis of SLE in the older population.

**Methods:** We conducted the current study to analyze characteristics and outcome of patients with late onset SLE in a Tunisian tertiary referral center, and to compare them with patients younger than 50 years at SLE diagnosis. This cross-sectional study consisted of 83 SLE patients (75 female and 8 male), who attended the Department of Internal Medicine in Monastir between 2006 and 2015.

**Results:** 12 patients were identified as having late-onset SLE, diagnosed at or over the age of 50 years. These patients were compared with a group of 71 young patients at SLE diagnosis. We compared clinical features, laboratory data, management and course.

Elderly patients less often presented with malar rash, photosensitivity, and nephropathy. Regarding laboratory features, older patients have a higher frequency of rheumatoid factor compared with young SLE patients. Whereas, leukopenia was less frequent in the older population

Deaths occurred more frequently in late-onset SLE (p = 0.02); the main causes of death were infectious complications.

**Conclusion: Late-onset SLE is characterized by a lower severity of disease. The poorer prognosis of late-onset SLE contrasting with the lower severity of the disease seems to result mainly from the consequences of aging.**

**Key words:** Late onset, early onset, systemic lupus erythematosus; elderly

## Introduction

Systemic lupus Erythematosus (SLE) is an autoimmune disease with wide clinical features. The diagnosis of SLE is based on American College of Rheumatology (ACR) and SLICC criteria. SLE affects usually young adults. Few reports studied SLE in the elderly [1,2,3,4].

We enrolled a retrospective study to establish the epidemiological, clinical and laboratory profile, therapeutic and prognostic aspects and course disease of late onset SLE. The aim of our study is to compare the different presentations of SLE in old and young population.

## Patients and Method

We conducted a cross-sectional study that consisted of 83 SLE patients (75 female and 8 male), who attended the Department of Internal Medicine in Monastir University Hospital between 2006 and 2015. All patients met at least 4 criteria of the American College of Rheumatology (ACR/SLICC) revised criteria of SLE [5]. Patients with drug-induced SLE or pure cutaneous lupus were excluded. Patients with diagnosis of SLE at the age of 50 or later were classified as the "late onset" lupus, all patients aged younger than 50 years at SLE diagnosis, constituted the ''early-onset'' SLE group. All patients underwent a physical examination. In addition to routine laboratory investigations including tests were conducted for rheumatoid factor, antinuclear antibody (indirect immunofluorescence method with HEp-2 cells as substrate), complements 3 and 4, anticardiolipin antibodies, and lupus anticoagulant. Other several examinations such as an echogram and computerized tomography were performed, if needed. The following items were recorded: age at the onset of the disease, sex, date of diagnosis, clinical and laboratory manifestations and the status at the last visit. Organ involvement not related to SLE was excluded. The SLE disease activity index (SLEDAI) was used to measure the severity of disease at the time of diagnosis [6].

**Statistical analysis:** The data were analyzed statistically using SPSS software for Windows (version 18) to compare the epidemiological, clinical and laboratory parameters in different groups. A Fisher exact test was performed for qualitative variables, and a Student t-test for comparison of quantitative variables. Statistical significance was defined as $p \leq 0.05$.

## Results

From the total of 83 (75 female and 8 male) patients included in this study, 12 patients were identified as having late-onset SLE; mean age at SLE diagnosis was 58.4 ± 5.8 years (range, 51-72 yr.). The early onset control group n= 71 (mean age 31.8 ± 10 years, range 16-49). There was no significant difference in gender distribution among the groups (Table 1).

The duration between disease onset and diagnosis was 26 months (range, 11-48) for the old patients, compared with 10 months (range, 3-24) in the younger SLE group. This difference was statistically significant (p=0.01). Mean number of revised ACR criteria was smaller for the older population compared to the younger one (4.9 ± 0.6 vs 6.8± 0.9; p = 0.04),

Clinical manifestations at diagnosis in both groups are summarized in Table 2 (next page).

In the Late onset group lupus showed a significantly less frequency of malar rash (33.3% vs 70.4 %; p=0.01), photosensitivity (41.6% vs 73.2 %; p=0.02) and lupus nephritis (8.3 % vs 36.6 %; p= 0.04) compared with the early-onset group.

Table 1 : Demographic characteristics of SLE in late and early onset patients

| | Late onset (n=12) | Early onset (n = 71) | p value |
|---|---|---|---|
| Mean age (years) | 58.4 ± 5.8 | 31.8 ± 10 | -- |
| Sex-ratio F/M | 11/1 | 64/7 | 0,86 |
| Delay diagnosis* | 26±9.4 | 10±6.4 | 0,01 |
| SLEDAI | 8.1 | 9.4 | 0.5 |
| ACR Criteria | 4.9 ± 0.6 | 6.8± 0.9 | 0.04 |
| Mean years of follow up | 6.8 ± 5.2 | 9.5 ± 5.8 | 0,02 |

*delay between SLE onset and diagnosis, F/M: female/male

**Table 2 : Clinical manifestations at the diagnosis of SLE in late and early onset patients**

| | Late onset, n=12 No. of patients (%) | Early onset, n= 71 No. of patients (%) | p value |
|---|---|---|---|
| Fever | 4 (33,3) | 19 (26.7) | NS |
| Weight loss | 3 (25) | 20 (28) | NS |
| Malar rash | 4 (33,3) | 50 (70,4) | 0,01 |
| Photosensitivity | 5 (41,6) | 52 (73,2) | 0.02 |
| Mucosal ulcer | 1 (8,3) | 11 (15,4) | NS |
| Discoid lupus | 2 (16,6) | 8 (11,2) | NS |
| Alopecia/hair loss | 2 (16,6) | 9 (12,6) | NS |
| Arthritis | 8 (66) | 48 (67,6) | NS |
| Lupus nephritis* | 1 (8.3) | 26 (36.6) | 0,04 |
| Pericarditis | 5 (41,6) | 16 (22,5) | NS |
| Pleuritis | 3 (25) | 16 (22,5) | NS |
| Lung involvement | 5 (41,6) | 19 (26,7) | NS |
| Neurological involvement | 3 (25) | 16 (22,5) | NS |
| Ocular involvement | 3 (25) | 9 (12,6) | NS |
| Thrombosis | 4 (33,3) | 11 (15,4) | NS |
| Pulmonary embolism | 1 (8,3) | 4 (5,6) | NS |

*Proliferative glomerulonephritis (WHO class II, III or IV)

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## Laboratory Findings

Regarding laboratory findings at the time of onset of SLE (Table 3), the late-onset group was characterized by a lower frequency of leukopenia (25% vs 55 %; p=0.05). The other hematological disorders (lymphopenia, hemolysis anemia and thrombocytemia) were slightly less frequent compared to the early-onset group, but differences were not significant. Whereas rheumatoid factor positivity occurred more frequently in the elderly. No significant differences were found for anti-dsDNA, anti-Sm, and antiphospholipid antibodies

Table 4 summarizes the main concomitant auto-immune diseases associated to SLE. The late onset of lupus was associated with increased incidence of rheumatoid arthritis (33.3 % vs 7%; p=0.007). APS and Sjogren syndrome were slightly more frequent compared to the early-onset group.

## Therapy and Course

High-dose corticosteroids >1mg/kg/day (42.2 % vs. 18 %; p = 0.05) and hydroxychloroquine (92.9 % vs 75 %; p = 0.007) were less frequently used in the late-onset group.

The late onset of lupus was associated with increased incidence of hypertension, diabetes, osteoporosis and infections. At the time of analysis, 3 patients had died in the late onset group of which 3 were infections, and only 4 patients in the early onset group (p=0.02). None of our late-onset SLE patients died of SLE flare.

## Discussion

SLE has been considered to be a disease of young women. Late onset SLE is less frequent. From the total of 83 SLE patients in this study, twelve late-onset SLE patients, that is, diagnosed at or over the age of 50 years, represented 14.4% of all patients with SLE followed in our department. Different incidences have been reported (3.6-20 %). This may be related to the lack of strict definition of late-onset SLE. The cutoff age used most frequently is 50 years at disease or diagnosis [1,2, 7,8]. The female-to male ratio declines but not significantly with age in our study. In most of the literature data, the female to male ratio was significantly lower in the late onset group [7]. This probably reflects the relationship between SLE and estrogen status [8-9]. Oestrogens have multiple immunomodulatory effects. There was a significant delay in the diagnosis of SLE in older ages. This longer interval may be related to the atypical presentation

**Table 3: Laboratory findings of SLE in late and early onset patients**

| | Late onset, n=12 No. of patients (%) | Early onset, n= 71 No. of patients (%) | p value |
|---|---|---|---|
| Inflammatory syndrome | 8 (66,6) | 47 (66,1) | NS |
| Hemolysis Anemia | 0 (0) | 3 (4,2) | NS |
| Leukopenia<4000/mm3 | 3 (25) | 39 (55) | 0,05 |
| Lymphopenia | 8 (66,6) | 58 (81,6) | NS |
| Thrombocytopenia* | 1 (8,3) | 9 (14,5) | NS |
| Hypocomplementemia | 3 (25) | 17 (23,9) | NS |
| Antinuclear antibodies | 12 (100) | 65 (91,5) | NS |
| Anti-DNA antibodies | 9 (75) | 57 (80,2) | NS |
| Rheumatoid factor | 4 (33,3) | 12 (16.9) | NS |
| Anti-Sm antibodies | 2 (16,6) | 26 (48,1) | NS |
| Anti-SSA antibodies | 5 (41,6) | 35 (63,6) | NS |
| Anti-SSB antibodies | 4 (33,3) | 23 (41,8) | NS |
| Anti-RNP antibodies | 2 (16,6) | 20 (37,7) | NS |
| Anticardiolipin antibodies IgG | 4 (33,3) | 24 (35,2) | NS |
| Anti- β2GPI | 4 (33,3) | 13 (18) | NS |

**Table 4 : Autoimmune diseases associated with SLE in late and early onset patients**

| | Late onset, n=12 No. of patients (%) | Early onset, n= 71 No. of patients (%) | p value |
|---|---|---|---|
| Rheumatoid arthritis | 4 (33,3) | 5 (7) | 0,007 |
| APS * | 3 (25) | 13 (18,3) | NS |
| Sjögren's syndrome | 4 (33,3) | 18 (25,3) | NS |
| Thyroiditis | 2 (16,6) | 11 (15.4) | NS |
| Scleroderma | 0 (0) | 2 (2,8) | NS |

**Table 5 : Complications that occurred during follow up in late and early onset patients**

| | Late onset, n=12 No. of patients (%) | Early onset, n= 71 No. of patients (%) | p value |
|---|---|---|---|
| Osteoporosis | 7 (58,3) | 6 (8,4) | 0,0001 |
| Hypertension | 6 (50) | 7 (9.8) | 0,001 |
| Diabetes | 5 (41.6) | 11 (15,4) | 0,04 |
| Infection | 8 (66) | 18 (25.3) | 0,002 |
| Death | 3 (25) | 4 (5,6) | 0,02 |

of SLE in the older population. The disease seems to be mild with lower SLE criteria compared to the younger group. Similar results have been reported by many other studies [8, 10].

Age at disease onset of SLE influenced the clinical manifestations and serological laboratory findings. The result of our study shows that older population less often presented with malar rash, photosensitivity, and nephropathy. Most of the literature data segregating early and late onset group, show lower rates of nephritis, malar rash, photosensitivity, arthritis and less visceral involvement with more benign course in the older group, whereas, pulmonary involvement and serositis are more frequently observed [8, 10, 11, 12] . The results of our study support the concept that late onset lupus is less active in the older population [10- 16].

Regarding laboratory results, Leukopenia was significantly more frequent in the early onset group, whereas rheumatoid factor positivity was more frequent (33.3 % vs. 16.9 %) in old compared with young SLE patients, but the difference was not statistically significant, although no difference was found for other autoantibody frequencies (anti DNA, anti-Sm, anti-RNP, SSA and anti-SSB antibodies). Das Chagas Medeiros MM et al reported leuko/lymphopenia most frequently in late onset lupus but Appenzeller et al. reported hemolytic anemia and thrombocytopenia most frequently in late-onset SLE. Lalani et al. found no difference between the groups when comparing lymphopenia and thrombocytopenia [15, 17-18].

It should be noted that, it is difficult to be conclusive with regard to autoantibody findings, since they were not measured in the same laboratory. For the rheumatoid factor, most publications agree in indicating its higher frequency in patients with late-onset SLE [7, 19, 20], However, it should be noted that this is probably related to the prevalence of increased rheumatoid factor positivity in the elderly population. The literature is rather inconsistent with regard to the predominant serological profile in early and late onset SLE. Our study found no differences in the autoantibody profile in the two groups. Boddaert J et al reported a higher frequency of anti-RNP, anti-Sm and rheumatoid factor with a lower frequency of hypocomplementemia in late-onset SLE [7]. Appenzeller et al. reported a lower frequency of anti-Ro in late-onset SLE, while Alonso MD et al reported a lower frequency of positive anti-DNA [17,21]. Lower positive rates of autoantibodies and less frequent hypocomplementaemia may indicate milder disease activity in older population [3, 21]. Some series reported a higher prevalence of positive anti-Ro/SSA and/or anti-La/SSB antibodies [22-25]. In the present investigation, there were no differences in the frequency of Sjogren's syndrome among the group of late-onset SLE and the group of early-onset SLE patients, but rheumatoid arthritis was more associated to late onset SLE. Another cohort reported a higher frequency of Sjogren's syndrome in their populations with late-onset SLE [19, 27]; Boddaert J et al in their pooled data analysis, reported that Sjögren's syndrome was more frequently present in the late-onset than in the early-onset SLE, although no difference was found for anti-SSA and anti-SSB antibodies [7]. The lack of a parallel increase in Sjogren's autoantibodies in these patients may reflect the aging process, in fact the sicca symptoms may be related to other more frequent causes of mucosal dryness in the elderly population, such as senile salivary gland atrophy, polypharmacy and chronic debilitating diseases [27, 28].

Hypertension, diabetes and osteoporosis were significantly more increased in the late onset group than in the early onset group in our cohort. The greater frequency of age related comorbidities in older population is due to aging and longer exposure to classical risk factors and use of corticosteroids [7,29]. In our cohort, the mortality rate was greater in the late onset group; infection caused death in 3 of 4 patients. The higher mortality in older patients contrasting with the lower severity of the disease, may be related to the fact that lupus at that age may be complicated by co-morbidities and increased risk of toxicities from immunosuppressive drugs usually used.

In our study glucocorticoids, hydroxychloroquine use was less frequent in the late-onset group compared to the early-onset group.

Our study has some limitations. First the validity is limited by the retrospective design; the collection of the data may have underestimated the frequency of some clinical manifestations. Second, the size of our sample is quite small; it is representative only for the population of the center of Tunisia. We also acknowledge that there may have been changes to medications or co-morbidities outside our hospital.

In summary, this study in the Tunisian SLE population showed that age affects the expression of SLE. Late onset SLE has a milder disease compared with early onset group, characterized by less renal, photosensitivity and malar rash. However, the mortality rate of older population is higher. Co-morbidities such as hypertension, diabetes and osteoporosis were significantly more frequent in late-onset SLE.

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# Inherited Ataxias: An Overview

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## Introduction

The hereditary ataxias are a heterogeneous group of diseases. Most attempts at classification have been based on pathologic findings and are not always useful for the clinicians. Many of these disorders are multisystem degeneration in which the underlying biochemical or other defect is usually unknown. The pathophysiology is correspondingly poorly understood. Hereditary ataxia can be divided into the hereditary congenital ataxia, the ataxia linked with metabolic disorder, and early onset ataxia of unknown etiology (1) (Table 1). Primary care physicians should be aware of the differential diagnosis of hereditary ataxia when faced with ataxia of unknown etiology.

**Table 1. Classification of Hereditary Ataxia**

| |
|---|
| **I. Congenital Cerebellar Ataxia** |
| **II. Ataxia associated with metabolic disorders** |
|    a. Intermittent ataxia syndromes |
|    b. Progressive unremitting ataxia syndromes |
|    c. Ataxia disorders associated with defective DNA repairs |
| **III. Progressive ataxia disorders of unknown etiology.** |
|    a. Early - onset cerebellar ataxia (onset usually before age 20) |
|    b. Late - onset cerebellar ataxia (onset usually after age 20) |

## Classification

The degenerative cerebellar and spinocerebellar disorders are a complex group of diseases, most of which are genetically determined. Tremendous confusion exists in classifying degenerative disorders causing ataxia, and there is no universally accepted system, these disorders can be divided into two main groups, depending on whether onset of symptoms is before or after the age of 20 years. Most of the early onset are autosomal recessive, and the later onset ones autosomal dominant (2). Most of these disorders are multisystem degenerations in which the underlying biochemical or other defect is usually unknown; the pathophysiology is correspondingly poorly understood. The differential diagnosis of ataxia is important since some of them are treatable if detected early. The discussion will concentrate on congenital cerebellar ataxia, ataxia associated with metabolic disorders and progressive ataxia of unknown etiology.

### I. Congenital Cerebellar Ataxia

Non-progressive rare type of ataxia: Cerebellar dysfunction usually starts in infancy with abnormal motor development, and hypotonic. Other symptoms includes nystagmus, intention tremor, feeding problems, marked truncal ataxia, delay in ability to sit, and stand, and variable degree of mental retardation and spasticity (2). Classification of the congenital cerebellar syndromes are shown in Table 2. The inheritance pattern of the majority of these syndromes are of autosomal recessive, with a 25% recurrence risk to subsequent siblings.

**Table 2: Congenital Cerebellar Ataxias**

| |
|---|
| **Congenital ataxia with mental retardation and spasticity**<br>　(includes pontoneocerebellar hypoplasia)<br><br>**Congenital ataxia + /- mental retardation**<br>　(includes granule cell hypoplasia)<br><br>**Congenital ataxia with mental retardation, episodic hypernea, and abnormal eye movements**<br>　(Joubert's syndrome)<br><br>**Congenital ataxia with partial aniridia and mental retardation**<br>　(Gillespie's syndrome)<br><br>**Dysequilibrium syndrome**<br><br>**X-linked recessive ataxia with mental retardation and spasticity**<br>　(Paine's syndrome) |

## II. Ataxia Associated with Metabolic Disorders

Ataxia secondary to metabolic disorder may lead to either persistent progressive ataxia or to intermittent ataxia. The underlying pathophysiology is usually accumulation of neurotoxic substances such as ammonia. (Table 3).

**Table 3: Ataxia Associated with Metabolic Disorders**

| |
|---|
| **INTERMITTENT ATAXIA SYNDROMES**<br><br>**With hyperammonia**<br>**Aminoacidurias**<br>**Disorders of pyruvate and lactate metabolism**<br>**Progressive Unremitting Ataxic Syndromes**<br>　Abeta-and hypobetalipoproteinemia<br>　Isolated vitamin E deficiency<br>　Hexosaminidase deficiency *<br>　Cholestanolosis *<br>　Leukodystrophies * (metachromatic, late-onset globoid cell, adrenoleukomyeloneuropathy)<br>　Mitochondrial encephalomyopathies *<br>　Wilson's disease<br>　Ceroid lipofuscinosis*<br>　Sialidosis<br>　Sphingomyelin storage disorders*<br>**Ataxia Disorders Associated with Defective DNA Repair**<br>　Ataxia - telangiectasia<br>　Xeroderma pigmentosum<br>　Cockayne's syndrome |

\* Ataxia may not be prominent feature.

### A. Intermittent Ataxia Syndrome

The hyperammonemias are usually caused by deficiencies of urea cycle enzymes and are of autosomal recessive inheritance, with the exception of ornithine - Transcarbamylase deficiency, which is X-linked. The clinical manifestations include intermittent ataxia, dysarthria, vomiting, headache ptosis, involuntary movements, confusion seiuzures and mental retardation. These episodes are usually precipitated by intercurrent illness and high protein load (3).

Ornithine Transcarbamylase deficiency is the most common urea cycle enzyme deficiency; it is X-linked. Antenatal diagnosis is available for this deficiency and for some of the other hyperammonemias. Protein restriction is the treatment of choice in addition to intravenous fluid administration during acute episodes of neurologic dysfunction. Aminoacidurias usually have similar manifestations as that of the hyperammonemias. Inheritance is autosomal recessive and treatment consists of restricting the intake of branched chain amino acids through special diets (4). Pyruvate dehydrogenase disorders are heterogeneous and rare. When the level of enzyme activity is between 35 and 50% of normal a syndrome of intermittent ataxia and choreathetosis occur during febrile illness and mild cerebellar dysfunction between episodes (5). Severe lactic acidosis occurs when the activity of this enzyme is below 15% of normal (5). A ketogenic diet may slow progression of the disease.

## B. Progressive Unremitting Ataxia Syndrome

A number of hereditary metabolic ataxia lead to progressive unremitting syndrome (Table 3). Most of them are autosomal recessive disorders. Abnormal function of apolipoprotein B, the carrier of lipid from the intestinal cells to plasma, lead to Abetalipoproteinemia. The most prominent features of this disease include ataxia, areflexia, loss of vibration and position sense, and pigmentary retinopathy. Symptoms of fat malabsorption are often mild and may be overlooked. Serum cholesterol level is low and serum vitamin E concentrations are low or undetectable from birth and this deficiency may be the cause of neurologic disorders. Onset of symptoms is usually in the second decade of life. Large doses of vitamin E results in improvement or stabilization of symptoms or may prevent the development of neurological symptoms. Retinopathy may be related to deficiency of both vitamins A & E (6).

Isolated cases of vitamin E deficiency with no evidence of hypolipidemia or generalized fat absorption can occur (6). This is caused by a specific defect of vitamin E absorption which is inherited as an autosomal recessive trait. Adequate vitamin E therapy usually halts the progression of neurologic symptoms. Other disorders associated with ataxia include adrenoleukomyeloneuropathy, the sphingomyelin lipidoses (combined with supranuclear gaze palsy and dementia), metachromatic leukodystrophy, galactosylceramide lipidoses (Krabbe's disease), and the hexominidase deficiencies (7). A rare autosomal recessive disorder in cholestanolosis is caused by defective bile salt metabolism. This disease is characterized by ataxia, dementia, spasticity, peripheral neuropathy, cataract, and xanthomas in the second decade of life. Neurological function usually improves with treatment with chenodeoxycholic acid (8). Mitochondrial myopathies consist of various phenotypes with late onset ataxic disorders associated with such features as deafness, dementia, ataxia, peripheral neuropathy and myoclonus (9). Ataxia is an important albeit pathognomic finding in the poorly glycosylated glycoprotein syndrome.

## C. Ataxia Disorders Associated With DNA Repair

Ataxia - telangiectasia is the most common ataxic disorder associated with defective DNA repair. It is a multisystem, autosomal recessive disorder. It usually starts with recurrent sinopulmonary infections. Telangiectasis appear on the skin between 3 & 6 years. Ataxia is truncal and is progressive. Other symptoms include dysarthria, tremor, ophthalmologic and insulin resistance. Immunologic abnormalities include reduced level of secretary IgA serum IgE and abnormalities in cellular immunity. The cause of death is usually recurrent infections or malignant tumors. Helpful diagnostic clues are elevation of serum and fetoproteins (10).

A heterogenous disorder consisting of at least six genetically distinct autosomal recessive diseases is Xeroderma Pigmentosum. It is caused by reduced capacity to perform excision repair of DNA damaged by ultraviolet light and of some carcinoembryonic antigen. The disorders manifest mainly with skin photosensitivity, skin diseases malignancies and in three forms of the disease neurologic dysfunction occurs (11). In about half of the patients with neurologic disease, ataxia and hyporeflexia occur with mental retardation, deafness, involuntary movements and spasticity being more common.

## III. Progressive Ataxia Disorders of Unknown Etiology

These can be divided into two main groups, depending on whether onset of symptoms is before or after the age of 20 years. Most of the early onset disorders are autosomal recessive, and the later onset ones autosomal dominant (2).

## A. Early Onset Cerebellar Ataxia

### Friedreich's ataxia (FA)
Friedreich's ataxia is the most common of the early onset ataxia. It is one of the best defined and most common forms of hereditary ataxias of unknown etiology(1,2). In some large case series it comprises about 50% of the hereditary ataxia (2,12). It is transmitted in an autosomal recessive manner, and usually appearing in childhood or in adolescence but rarely in old age (13). The disease usually progresses slowly without remission, affecting both the central and peripheral nervous system (13-14). The most frequent first symptom is ataxia of gait.

The epidemiology of Friedreich's ataxia is perplexing. The clinical features and diagnostic criteria were defined by the Quebec Cooperative Study of Friedreich's Ataxia (QCSFA) (15) and by Harding (1,2 ) (Table 4). Both authors regarded recessive inheritance, progressive ataxia of limbs and gait and lower limb areflexia as obligatory criteria. The onset, according to the QCSFA and Harding (2, 15), should never occur after the age of 20 years, and always before 25, according to Harding (2). A recent case was reported in the literature where symptoms started at a later stage (16). Dysarthria, decreased lower limb deep sensation and weakness, obligatory signs for the QCSFA, are not considered essential for an early diagnosis by Harding (2). The diagnosis is made essentially on clinical grounds, CT scan of the brain may show mild cerebellar atrophy.

The prevalence is known only for some populations (12, 17-20). The range is from 0.6 to 1.4/100,000 population. The incidence has been estimated to be approximately 1-2/100,000 (17, 21).

Friedreich's ataxia is characterized by degeneration of the spinocerebellar pathways, the dorsolateral columns, and the dentate nuclei (1). There are few changes in the cerebellar cortex itself (1). The cerebrospinal fluid is usually normal and the CT scan of the brain is either normal or shows mild cerebellar atrophy. The primary clinical signs include ataxia, most marked in the lower limbs and often accompanied by dysarthria; nystagmus is usually present in 70% along with skeletal-muscle weakness (22). Optic atrophy and retinal pigmentation is usually present. Pes cavus and scoliosis almost always develop (23). Death is usually sudden and may be secondary to cardiac arrythmias (22). Cardiac involvement is frequent occurring in some 50% to 90% of cases (24); most commonly concentric hypertrophic cardiomyopathy is found (24,25).

Multiple studies have shown that the small coronary arteries are abnormal in patients who have cardiac disease and Friedreich's ataxia (26,27). The functional significance of this has been chal-

**Table 4: Friedreich's Ataxia : Diagnostic Criteria**

**Essential Criteria for Diagnosis: Present in More than 95% of Cases**
Autosomal recessive inheritance
Age at onset of symptoms before 25 years
Progressive limb and gate ataxia
Absent knee and ankle jerks
Extensor plantar responses
Motor nerve conduction velocity > 40m/s in upper limbs

**Small or undetectable sensory action potentials Additional Criteria, Not Essential for Diagnosis: Present in More than 65% of Cases**
Dysarthria*
Pyramidal weakness of lower limbs
Absent reflexes in upper limbs*
Distal loss of joint position and vibration sense in lower limbs*
Scoliosis
Abnormal electrocardiogram

**Other Features Present in 50% of cases or less**
Nystagmus
Optic atrophy
Deafness
Distal weakness and wasting
Pes cavus
Diabetes

lenged by Hewer (26). Biller et al. (27) reported a prevalence of 1.5% of cerebral infarction in 131 patients. It occurred in half of the patients who developed atrial fibrillation or atrial flutter with underlying symptomatic cardiomyopathy (27). Speech disorder is common in FA (28).

Electrophysiological and pathological studies suggest that axon degeneration and secondary demyelination occur in peripheral sensory nerves (29).

Electrophysiological evaluation of FA patients usually includes determination of motor and sensory conduction velocities (MCV, SCV) and multimodal evoked potentials (30). The degeneration of peripheral sensory and somatosensory pathway is usually measured by using nerve conduction studies and somatosensory evoked potential (SEPs) and brain-stem auditory evoked potentials (BAEPs) and the blink reflex (30).

Biochemical alterations observed in this disease include a reduced insulin receptor activity which leads to an insulin resistance state and a reduced glucose tolerance in about 40% of patients (31). Several lipid abnormalities have been noted as well, including a striking reduction in linoleic acid (32), low cholesterol levels with a total cholesterol reduction in serum and in the LDL and HDL fractions are described (32).

The results of therapeutic trials in Friedreich's ataxia with a number of drugs, including choline chloride, lecithine, physostigmine, y-vinyl aminobutyric acid, 5-hydroxytrptophan, benserazide,and thyrotropin releasing hormone, have been inconsistent or unconfirmed in terms of producing functional neurologic improvement.(2)

Amantadine hydrochloride (AH) is known to stimulate dopamine release (33). The use of AH in FA and OPCA was recently tested (34). Both studies revealed an improvement in reaction time (RT) and movement time (MT).

So there is no treatment known to influence the slowly deteriorating disease course. In order to minimize disability and prolong ambulation, strengthening and stretching exercises and functional retraining including aerobic endurance exercise are recommended (35).

***Early - Onset Cerebellar ataxia with Retained Tendon Reflexes***
The other early onset ataxia are listed in Table (5) . They are usually rare, with the exception of early onset cerebellar ataxia with retained reflexes, which occurs at a frequency about one quarter of that of FA, and is often confused with it, but is genetically distinct. The main clinical difference is that the tendon reflexes are normal or brisk in the disorder(36). It is important to distinguish between these two disorders, since the prognosis is better in the former, with patients losing the ability to walk on average 13 years later than in FA. In addition, severe skeletal deformity, heart disease, and diabetes do not occur (37).

***Cerebellar Ataxia with Hypogonadism***
The association of progressive ataxia with hypogonadotrophic hypogonadism is rare (2). Neurological symptoms usually develop in the third decade and hypogonadism is obvious at puberty. Neurological syndromes include dysarthria, nystagmus, progressive limb and gait ataxia, mental retardation, dementia deafness, choreoathetosis, retinopathy and sensory loss.

**Table 5: Early-Onset Ataxic Disorders of Unknown Etiology**

| |
|---|
| **Friedreich's ataxia** |
| **Early-onset cerebellar ataxia with** |
|   Hypogonadism |
|   Myoclonus (idiopathic Ramsay Hunt syndrome, progressive myoclonic ataxia) |
|   Pigmentary retinopathy |
|   optic atrophy + or - mental retardation |
|   cataract and mental retardation (Marinesco-Sjogren syndrome) |
|   Deafness |
|   Extrapyramidal features |
|   X-linked recessive spinocerebellar ataxia |

**Table 6: Late-Onset Ataxic Disorders Of Unknown Etiology**

| |
|---|
| **Autosomal dominant cerebellar ataxia (ADCA) with** |
|   Ophthalmoplegia, dementia, optic atrophy, extrapyramidal features and amyotrophy may include |
|   Machado- Joseph disease) |
|   (ADCA type I) |
| **ADCA with pigmentary retinopathy +/- Ophthalmoplegia and extrapyramidal features** (ADCA type II) |
| **Pure ADCA of later onset (after age 50)** (ADCA type III) |
| **Periodic ADCA** |
| **Other syndromes** |

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### Cerebellar Ataxia with myoclonus

The association of cerebellar ataxia and myoclonus, is often referred to as the Ramsay Hunt syndrome. This is a very heterogenous entity. Some of the identifiable causes include Baltic myoclonus, mitochondreal encephalomyopathy, and sialidosis (37). The rest of cases can be labelled as progressive myoclonic ataxia (37). Symptoms include the development of stimulus - sensitive myoclonus or generalized seizures at the end of the first decade of life. Ataxia and dysarthia develop a few years later with pyramidal signs in the limb. The myoclonic part of this syndrome may respond to clonazepan or valproate sodium with marked improvement in motor function.

### B. Late Onset Cerebellar Ataxia

These disorders have proved the most difficult and controversial in terms of classification (Table 6). The pathological findings are heterogenous reflecting huge clinical variations in the dominant ataxia (2).

### Autosomal Dominant Cerebellar Ataxia Type I (ADCA Type I).

The age of onset of symptoms in this syndrome ranges from 15 to 65 years but is most commonly in the third or fourth decade of life. Ataxia of gait is the most frequent presenting symptom; it usually involves the limbs and is invariably associated with dysarthria. Early onset usually predicts more progressive disability (38). Associated symptoms may include ophthalmoplegia, nystagmus, lid retraction and optic atrophy. Bulbar symptoms are common during the later stages of disorder and predispose the patient to respiratory infection. Other common symptoms include dementia, extrapyramidal signs, wasting and fasiculation of the face and tongue.

### Autosomal dominant cerebellar ataxia Type II (ADCA Type II).

This is clinically and genetically different from ADCA type I.

It is characterized in all families having retinopathy. The age at onset is earlier than that of ADCA type I, most commonly occurring between 15 and 35 (2,39).

### Autosomal Dominant Cerebellar Ataxia Type III

This is relatively pure cerebellar syndrome in which dementia, ocular or extrapyramidal features do not occur and onset of symptoms are usually after the age of 50 years(3). Nystagmus and pyramidal signs in the limbs are quite common.

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