

# INTRODUCTION

Amyloidosis are group of diseases that result from abnormal extracellular deposition of a particular protein, called amyloid, in various tissues of the body as insoluble fibrils causing progressive organ dysfunction . Amyloid protein can be deposited in a localized area and may not be harmful or only affect a single tissue of the body. This form of amyloidosis is called localized amyloidosis. While amyloidosis that affects tissues throughout the body is referred to as systemic amyloidosis, which can cause serious changes in virtually any organ of the body **(Pepys, 2006)**.

Types of amyloidosis are based on the precursor protein that form the amyloid fibrils and the distribution of its deposition, The involved organs are the heart, gastrointestinal tract , lung, medium sized blood vessels of all visceral organs , only the spleen appears to be devoid of B2microglobulin amyloid deposition . The kidney is affected in 50%-80% of individuals **(National Institute et al, 2009)**.

The major types of systemic amyloidosis are immunoglobulin (Ig) light chain , Ig heavy chain , familial or hereditary amyloidosis, Amyloid amyloidosis senile systemic amyloidosis, dialysis related amyloidosis. **( Noel et al, 1987)**.

Dialysis related amyloidosis (DRA) is one of the most serious complications associated with long term hemodialysis.It may be seen occasionally in patient with long standing renal failure who are not treated by dialysis **(Maeda ,2000)**.

Beta2 microglobulin was the major constituent protein of dialysis related amyloidosis,It is formed of a glycosylated polypeptide

consisting of 100 amino acid arranged in a single chain . the body produces 50 to 200 mg of Beta2 microglobulin and its normal serum level is 0.5 to 2.0 mg/L **(Gejyo et al,1985)**

The kidney is the only known major pathway for Beta2 microglobulin elimination which explain the accumulation of Beta2 microglobulin in patient with increase duration of uremia and dialysis **(Connors et al, 1985)**

Amyloidosis is chronic and usually progresses slowly over a number of years. The severity of the disease depends on which organs are affected by the amyloid deposits. Amyloidosis can be potentially life-threatening when the kidneys and heart are affected. **(Van – Ypersele et al, 1994)**

Early diagnosis is the key to managing the disease before it becomes fatal, DRA is relatively common in patients, especially older adults,and who have been on hemodialysis for more than 8 years **(Gertz et al, 1992)**

The clinical presentation of dialysis related amyloidosis is carpal tunnel syndrome,large and medium sized joint arthropathy which presents by arthralgia , decrease joint mobility , effusion incapacity,spondyloarthropathy which most frequently occur at the cervical spine and this may be asymptomatic or lead to mild spinal pain or may cause nerve root compression **(Deforges – Lasseur et al, 1993)**, bone cyst contain B2microglobulin amyloid deposits,intestinal obstruction , spontaneous tendon rupture , renal stones composed of B2microglobulin may occur , also myocardial dysfunction secondary to B2microglobulin deposits that become life-threatening **(Bardin et al ,1995)**

One of the common sign of kidney amyloidosis is the presence of proteinuria.

The diagnosis of amyloidosis requires histological demonstration of amyloid deposits , any tissue can be evaluated for congo red stain positivity , The likelihood of a missed diagnosis is lower with a kidney biopsy than with biopsies of other tissues because amyloid fibrils are visible by electron microscopy **(Balal et al ,2004)**

Light electron microscopy is the most direct method for identifying the amyloidogenic protein by mass spectrometry or amino acid sequencing of proteins that are extracted from the amyloid deposits , Computed tomographic scan and magnetic resonance imaging scan of bone will provide additional information of bone complications

Symptomatic treatment with steroid therapy, nonsteroidal anti-inflammatory drugs , Endoscopic and Surgical therapy including joint replacement and carpal tunnel release as necessary **(Zingraff et al ,1995)**

The use of high flux membrane in hemodialysis such as the polyslfone F60 or the polyacrylonitrite AN69 membrane allow to clear more or less marked amount of polypeptide and to remove large molecular weight solutes including B2microglobulin with better Biocomatibility than conventional unsubstituted cellulose **(Sanchez et al , 1993)**

Early renal transplantation is considered suitable for all patient before the development of established dialysis related amyloidosis **(Bardin et al ,1995).**