RENAL INVOLVEMENT AND PROGNOSTIC CRITERIA IN AMYLOIDOSIS

OSPEDALE GENERALE
"S. GIOVANNI CALIBITA" FATEBENEFRATELLI
ISOLA TIBERINA

M.G. Chiappini[1], T. Ammann[1], L. Muzi[1], E. Proietti[1], P. Pasqualetti[2], I. Simonelli[2], A. Grosso[1]

[1] Nephrology and Dialysis Unit FBF Hospital, Rome, Italy
[2] Medical Statistics e Information Technology, Fatebenefratelli Association for the Research, Rome, Italy

INTRODUCTION AND AIMS

Early diagnosis and supportive therapy are of primary importance in prognosis of patients with systemic amyloidosis. The effort increasing the awareness and the medical approach allowed to achieve earlier diagnosis in this rare condition. The purpose of this retrospective study was to improve the prognosis of patients with systemic amyloidosis identifying prognostic factors that have greater clinical relevance in their survival.

METHODS

Were studied 138 patients with amyloidosis: AL n=69 (M 45 F 24, mean age 63±12 years), AA n=14 (M 3 F 11, mean age 57±21 years), secondary to myeloma MA n=9 (M 5 F 4, average age 65+15 years), from TTR n=30 (M 19 F 11, average age 63 +11), from Apolipoprotein n=1 (M, age 51), of indeterminate type AX n=15 (M 9 F 6, mean age 63±9 years).

Were considered: age, type of amyloidosis, diagnostic criteria, the time between onset of disease and diagnosis, clinical signs and symptoms at onset and diagnosis, organs involved, nutritional state. Renal survival was identified with the start of dialysis. The involvement of an organ was based mainly on clinical consensus and the organs were divided into 5 groups: cardiac, renal, hepatic, neurological and articular. For all patients the diagnosis of amyloidosis was obtained with the positivity histological to Congo red in one or more organs. All patients were treated with specific therapy and for each patient was carried out a follow-up from onset of disease to death or until the last clinical control available.

CHARACTERISTICS OF PATIENTS

	AL	AA	MA	TTR	APO LIPOPROTEINA	AX	TOT
PATIENTS N	69 λ48 - κ16 altro 5	14	9	30	1	15	138
GENDER M/F	45/24	3/11	5/4	19/11	1/0	9/6	82/56
AGE YEARS	63±12	57±21	65±15	63±11	51	63±9	60±14

PARAMETERS

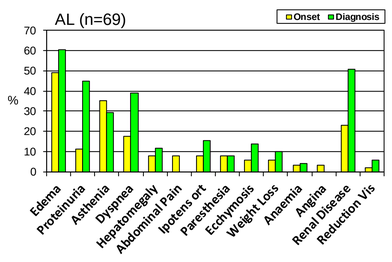
- GENDER
- AGE
- AMYLOIDOSIS TYPE
- TIME INTERVAL FROM ONSET TO DIAGNOSIS
- CLINICAL SIGNS AND SYMPTOMS AT ONSET
- CLINICAL SIGNS AND SYMPTOMS AT DIAGNOSIS
- IMMUNOHISTOCHEMICAL TESTING
- DIALYSIS YES/NO
- RENAL SCORE AT ONSET
- TREND RENAL INVOLVEMENT FROM ONSET TO FOLLOW-UP
- TREND GFR FROM ONSET TO FOLLOW-UP
- CARDIAC SCORE AT ONSET
- ORGANS INVOLVED AT ONSET, DIAGNOSIS, FOLLOW-UP
- Hb AT ONSET, DIAGNOSIS, FOLLOW-UP
- ALBUMIN AT ONSET, DIAGNOSIS, FOLLOW-UP

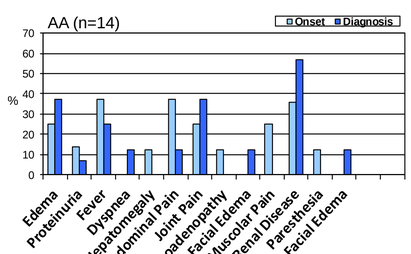
SCORE OF RENAL INVOLVEMENT
0 = ABSENT
1 = PROTEINURIA
2 = NEPHROTIC SYNDROME
3 = RENAL DISEASE

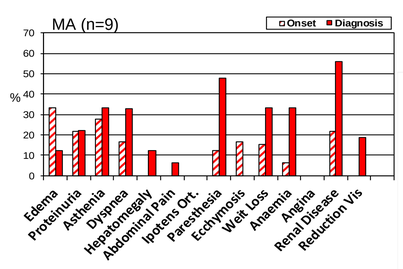
SCORE OF CARDIAC INVOLVEMENT
0 = ABSENT
1 = HYPERTROPHIC CARDIOPATHY
2 = DILATED CARDIOPATHY
3 = ARRHYTHMIAS
4 = ARRHYTHMIAS + HEART FAILURE

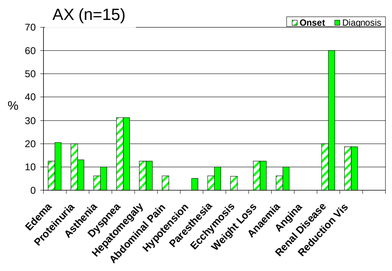
RESULTS

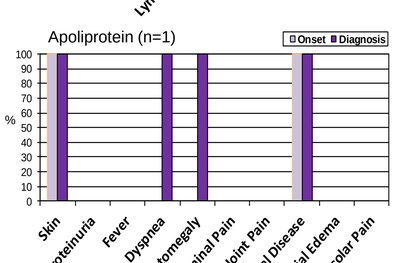
Clinical signs and symptoms onset and diagnosis

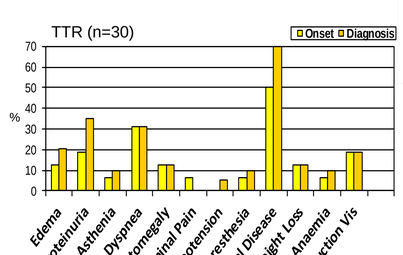




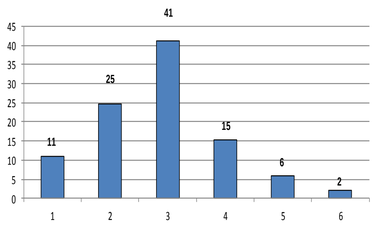




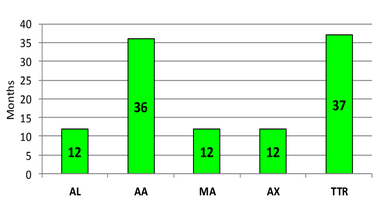




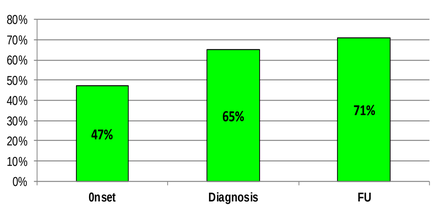
NUMBER ORGANS INVOLVED



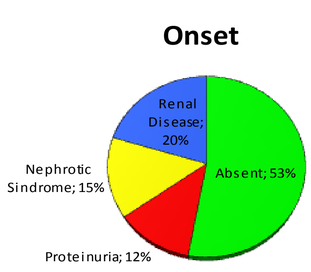
TIME ONSET-DIAGNOSIS (MEDIAN)

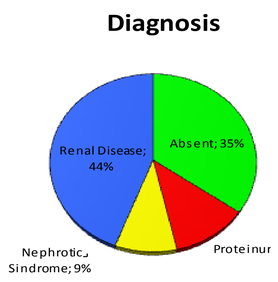


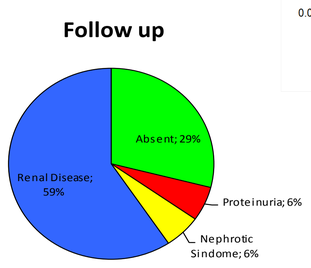
RENAL INVOLVEMENT



RENAL INVOLVEMENT=138







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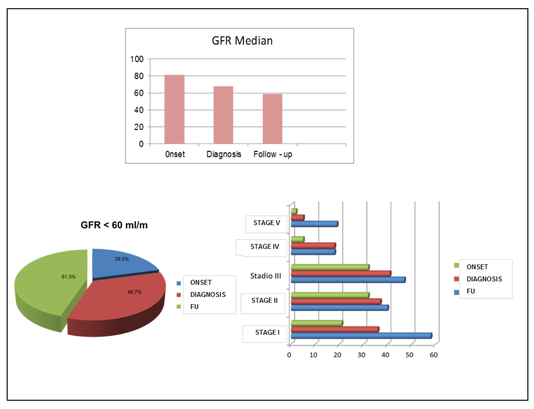
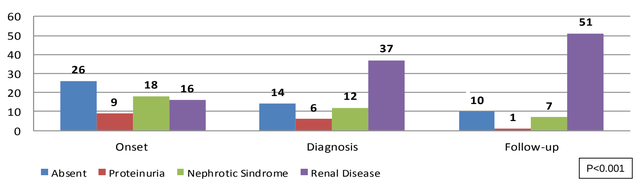
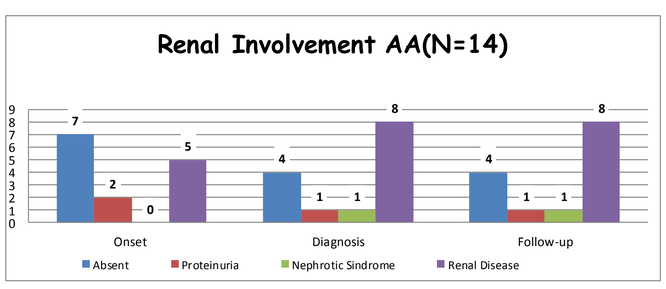


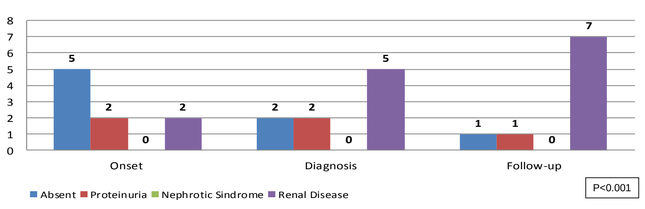
Renal Involvement AL (N=69)



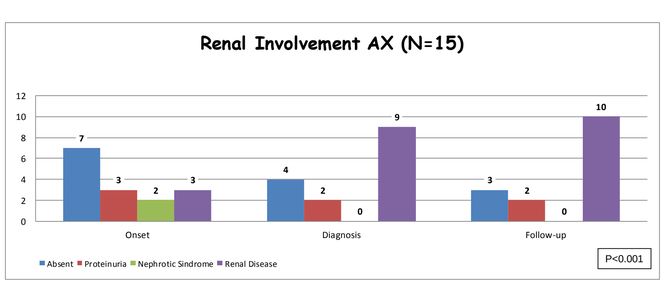
Renal Involvement AA(N=14)



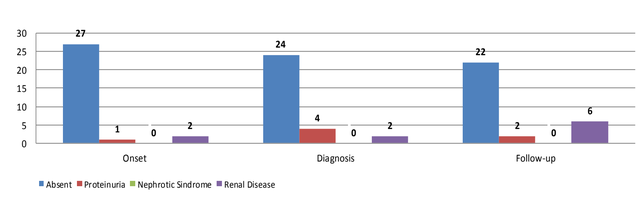
Renal Involvement MA (N=9)



Renal Involvement AX (N=15)



Renal Involvement TTR (N=30)



CARDIAC INVOLVEMENT N=138

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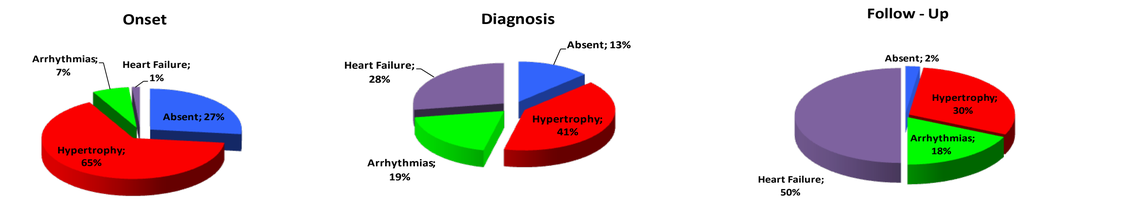
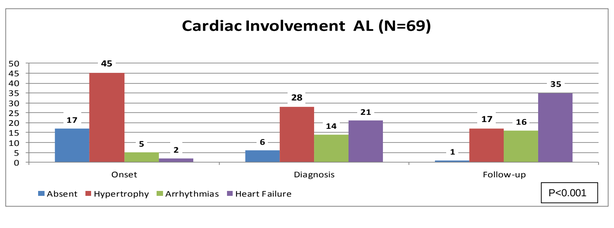
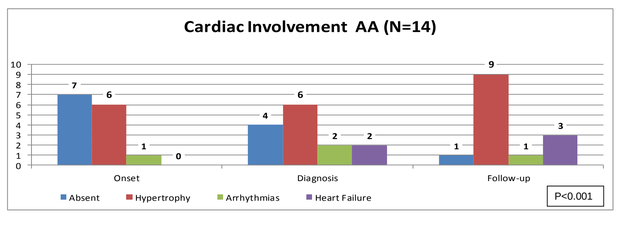
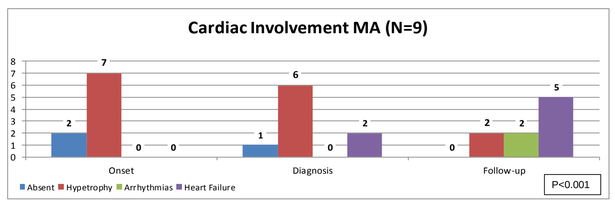
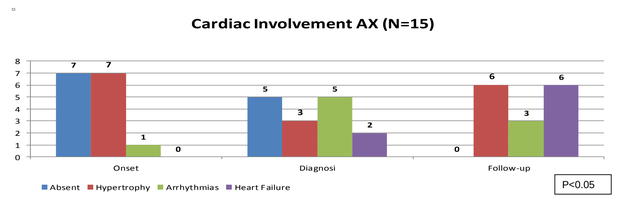
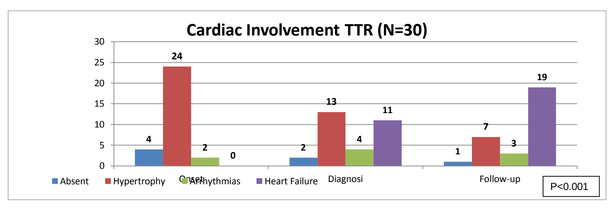


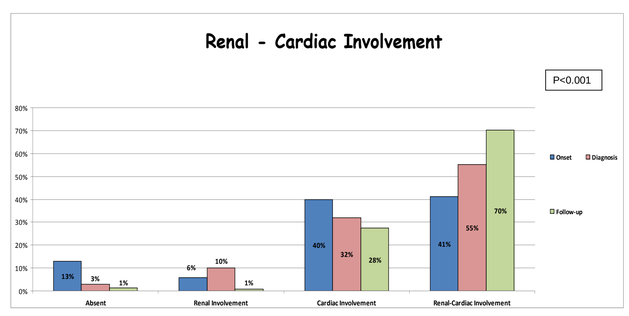






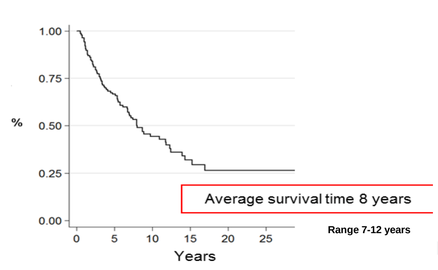




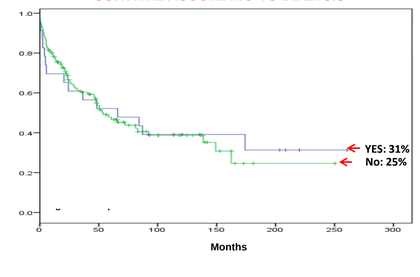


FOLLOW-UP (N=138)

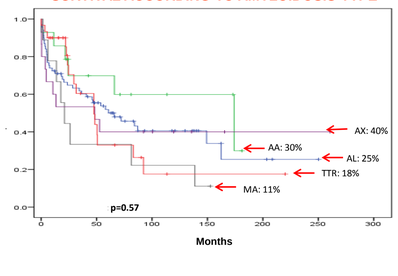
CUMULATIVE SURVIVAL (N=138)



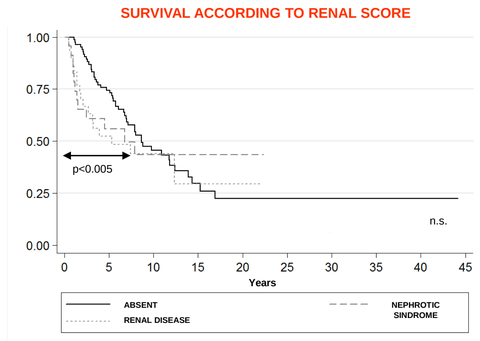
SURVIVAL ACCORDING TO DIALYSIS



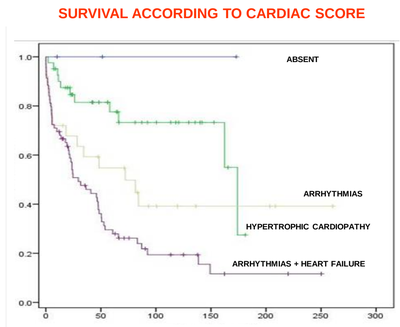
SURVIVAL ACCORDING TO AMYLOIDOSIS TYPE



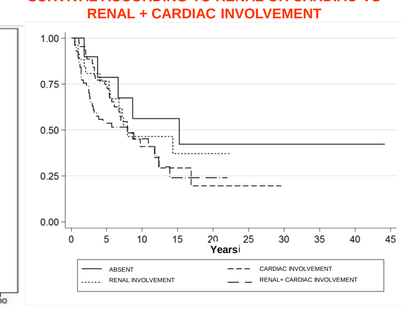
SURVIVAL ACCORDING TO RENAL SCORE



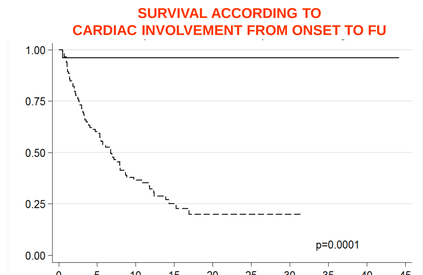
SURVIVAL ACCORDING TO CARDIAC SCORE



SURVIVAL ACCORDING TO RENAL OR CARDIAC VS RENAL + CARDIAC INVOLVEMENT



SURVIVAL ACCORDING TO CARDIAC INVOLVEMENT FROM ONSET TO FU



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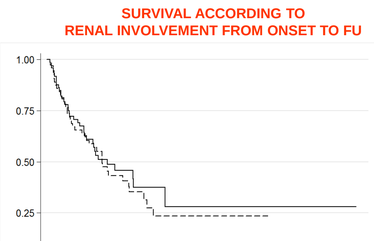
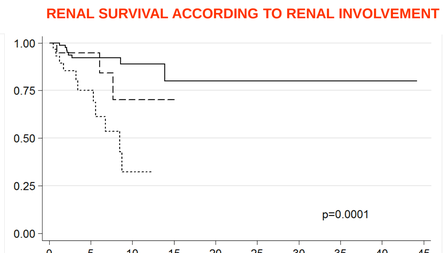


RENAL SURVIVAL ACCORDING TO RENAL INVOLVEMENT



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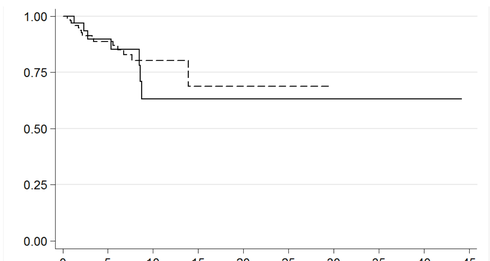
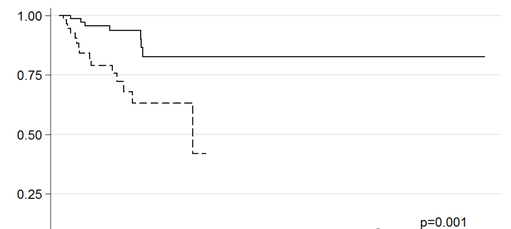


RENAL SURVIVAL ACCORDING TO RENAL OR CARDIAC VS RENAL+CARDIAC INVOLVEMENT



COX REGRESSION ANALYSIS

Predictors of mortality

ALBUMIN	p<0.001
AGE OF ONSET	p<0.001
DEGREE OF CARDIAC INVOLVEMENT AT ONSET	p<0.001
CARDIAC WORSENING FROM ONSET AT FOLLOW-UP	P<0.002
ORGANS INVOLVED	p<0.002

CONCLUSIONS

- The diagnosis of amyloidosis is late
- The renal and cardiac involvement they are present in a very elevated percentage of cases to the diagnosis
- New therapies seem to condition significantly the survival that results improved in comparison to the cases previous
- There is not any significant difference of the survival in comparison to the type of amyloidosis
- The degree of renal involvement influence significantly the prognosis within 10 years of follow .up, after the datum it is not more significant for the greater numerousness of the patients with amyloidosis TTR that almost exclusively showed a cardiac involvement
- The entry in dialysis doesn't seem to influence the prognosis of the patients
- The degree of cardiac involvement influence significantly highly the prognosis
- The degree of cardiac involvement influence in significant highly the prognosis
- The cardiac and renal involvement influence mostly the prognosis in comparison to the involvement of one of the two organs
- The analysis of cox has shown that albumin, age of onset, degree of cardiac involvement at onset, cardiac worsening from onset at follow-up, number of organs influence significantly the survival

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