

The Adult with Sickle Cell Disease

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Sickle Cell Disease

- Acute Complications:

Pain, Septicemia, CVA, Acute Chest Syndrome, Cholecystitis, Priapism, Splenic/Hepatic Sequestration, etc

- Chronic Complications:

Retinopathy, Restrictive lung disease/pulmonary hypertension, Left ventricular dysfunction, Cholestasis, Renal disease, Osteonecrosis, Leg ulcers, etc.

CHALLENGES in MANAGEMENT of the ADULT PATIENT

- Shorten the duration of acute complications
- Reduce the frequency of acute complications
- Reduce the frequency/severity of chronic complications
- Improve survival
- Enhance the Quality of Life

Annual Examination

- **H/O sickling related events:** Pain-acute & chronic, CVA, PNA/ACS (ICU), Hepatitis, Cholelithiasis, Priapism, Retinopathy, Pregnancy, Leg Ulcer, Hematuria/Infection, Bone infections/AVN, Transfusion
- **Lab:** CBC, Bun/Creatinine, Urine protein, Iron/Ferritin, Hepatitis markers, B12, Folate
- **X-ray:** Chest, Hips, Shoulders, T/L Spines
- **Other:** EKG, Echocardiogram, as needed

Outcome of Sickle Cell Anemia

- n = 1,056; 11,427 patient-yrs since 1959
- Males - 525, Females - 531

Follow-up started at

Age	n	%
< 1 years	190	18
< 10 years	222	21
< 20 years	169	16
> 20 years	475	45

Acute Clinical Events in Hb SS

Event	# of pts	% of pts	Incidence per 100 pt-yrs (mean)
Hospitalizations	805	76	79
Sickle related	766	73	67
Uncomplicated pain	739	70	57
2^o sickle pain	536	51	39
ACS	506	48	14
Hypersplenism	216	20	5
Bone infarct	160	15	3
Aplastic crisis	143	14	2
Trauma	140	13	2
Meningitis/Sepsis	128	12	3
CNS disorder	128	12	3
Dactylitis age < 4	41	4	3

Chronic Organ Damage in Hb SS

Organ	# of pts	% pts	Age at Dx (median)
Gall bladder	297	28.1	28
AVN	224	21.2	30
Lung	165	15.6	32
Leg ulcer	152	14.4	30
Priapism	71	13.5	29
Renal	122	11.6	37
CVA	116	11.0	20
Retina (III,IV)	92	8.7	30
Non-sickle	30	2.8	24
Composite	540	48.5	24
Mortality	232	22.0	31

Hb SS; Additional End-Organ Damage

Condition	n	Add'l n	Common Additional EOF
CLD	167	129 (77%)	GB-10%, LU-5%, AVN-5%
Renal	123	92 (75%)	GB-9%, LU-8%, AVN-8%
Retinopathy	92	68 (74%)	GB-15%, LU-9%, AVN-9%
AVN	224	165 (74%)	GB-21%, CLD-10%, LU-7%
Priapism	71	49 (69%)	GB-16%, AVN-12%, LU-6%
CVA	116	59 (51%)	AVN-10%, GB-8%
GB	297	184 (62%)	AVN-15%, LU-12%, CVA-7%
LU	152	81 (53%)	GB-15%, AVN-10%, CLD-7%

Pulmonary Disease (CSSCD)

- N = 318, > 18 y of age (mean 31 ± 10), 41 % male
- 90 % abnormal

Restrictive - 74 %

Obstructive - 1 %

Mixed O/R - 2 %

↓ D_LCO - 13 %

- Association between lung & kidney disease

Pulmonary Hypertension

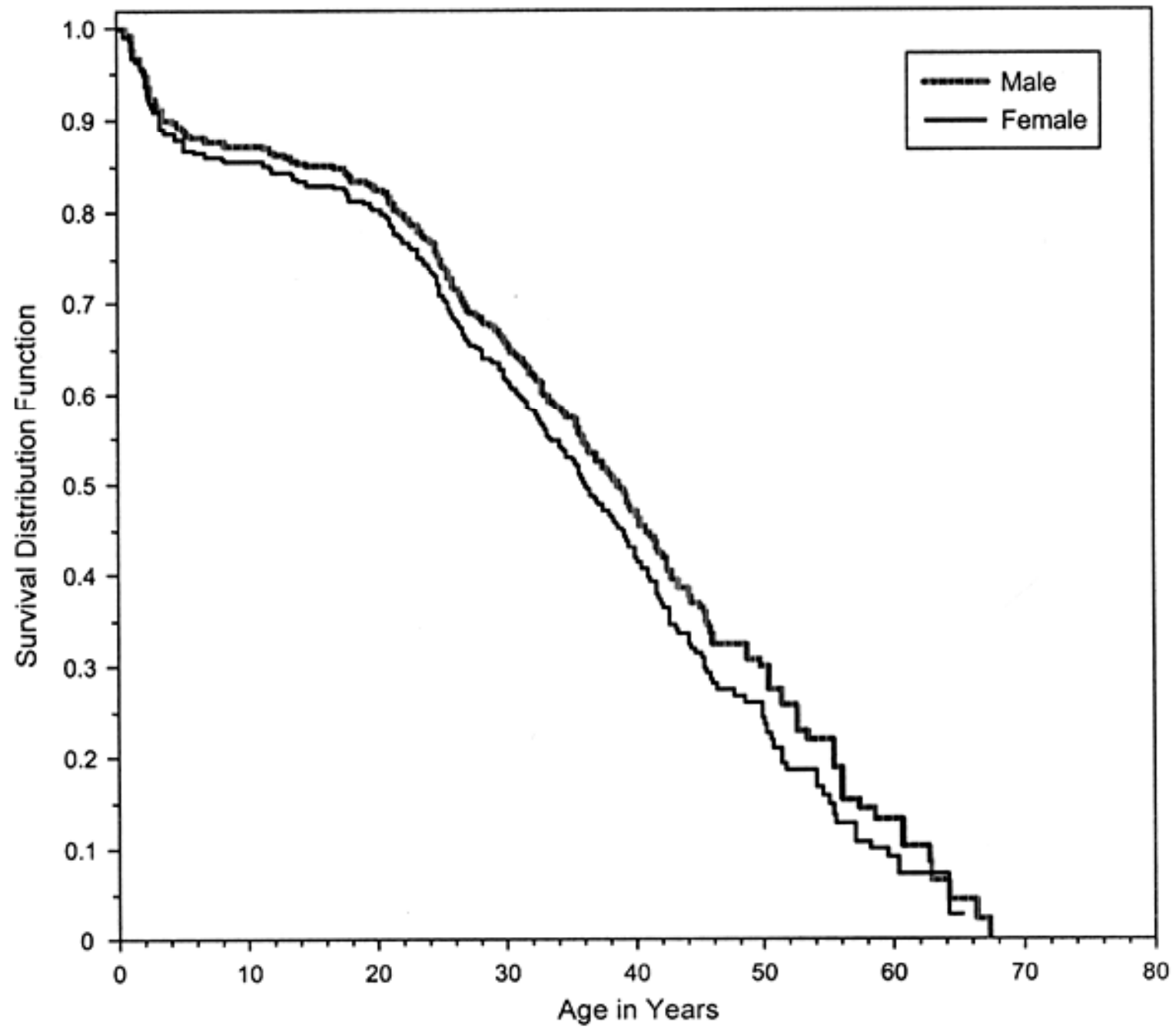
- Surveys with echocardiography reveal an incidence of ~10% in children and 35% in adults.
- Elevated pulmonary artery pressures are associated with shortened survival, pulmonary emboli/thrombosis, leg ulcers, priapism and increasing hemolysis

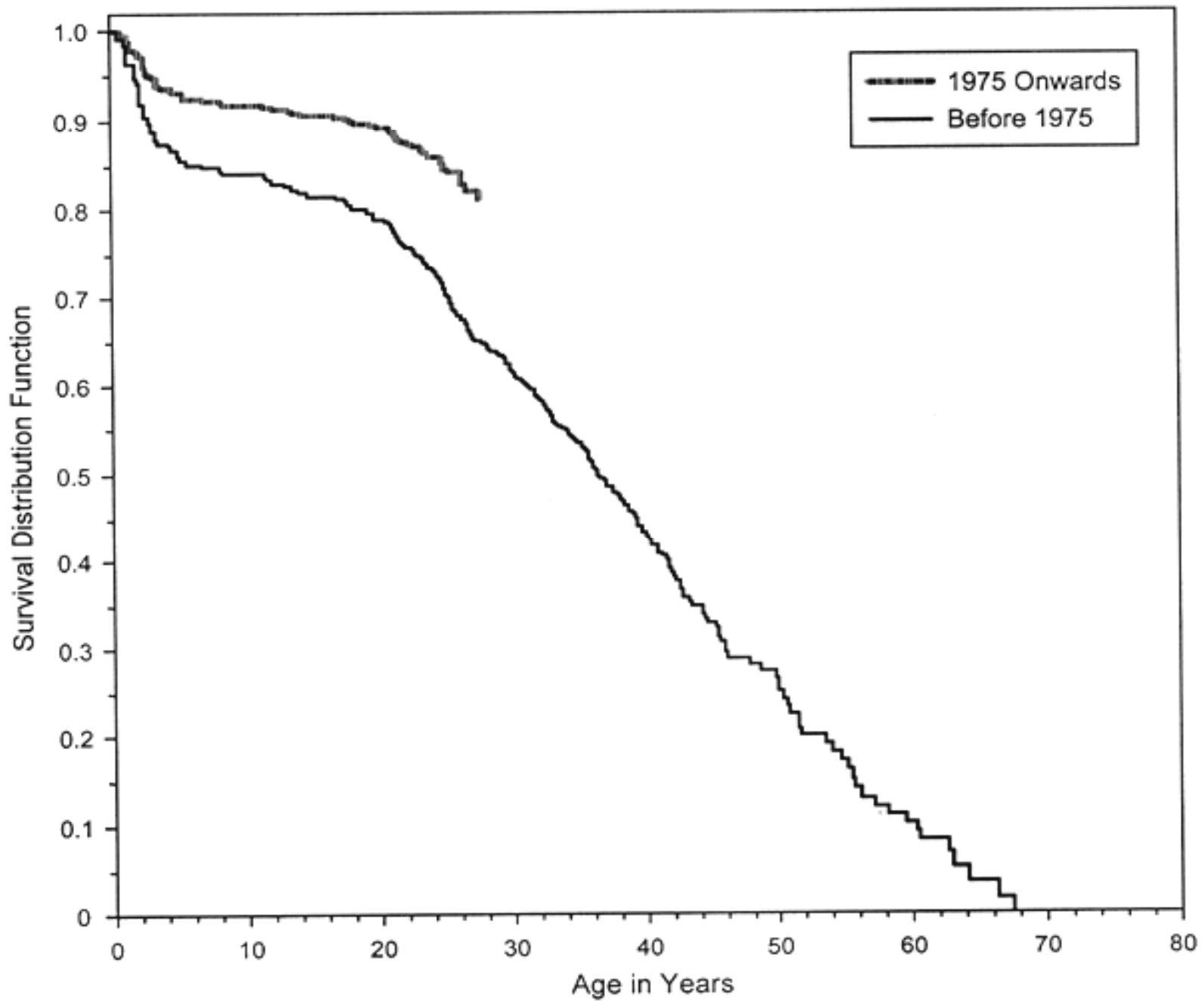
Priapism (CSSCD)

	p
CVA	0.53
AVN	0.006
ACS	< 0.001
Pain	< 0.001
↓ Anemia	< 0.001
↑ Bilirubin	< 0.001
↑ LDH	< 0.001
↑ Retic ct	< 0.001
↑ AST	0.005

Outcome in Hb SS: Mortality (%)

Cause	Age < 20 y, n=46	Age > 20 y, n=186
Infection	36	5
Hypersplenism	13	
End-organ failure	13	42
CNS	11	13
Trauma	9	8
ACS/MOF	7	11
Other	2	13
No Dx	5	8

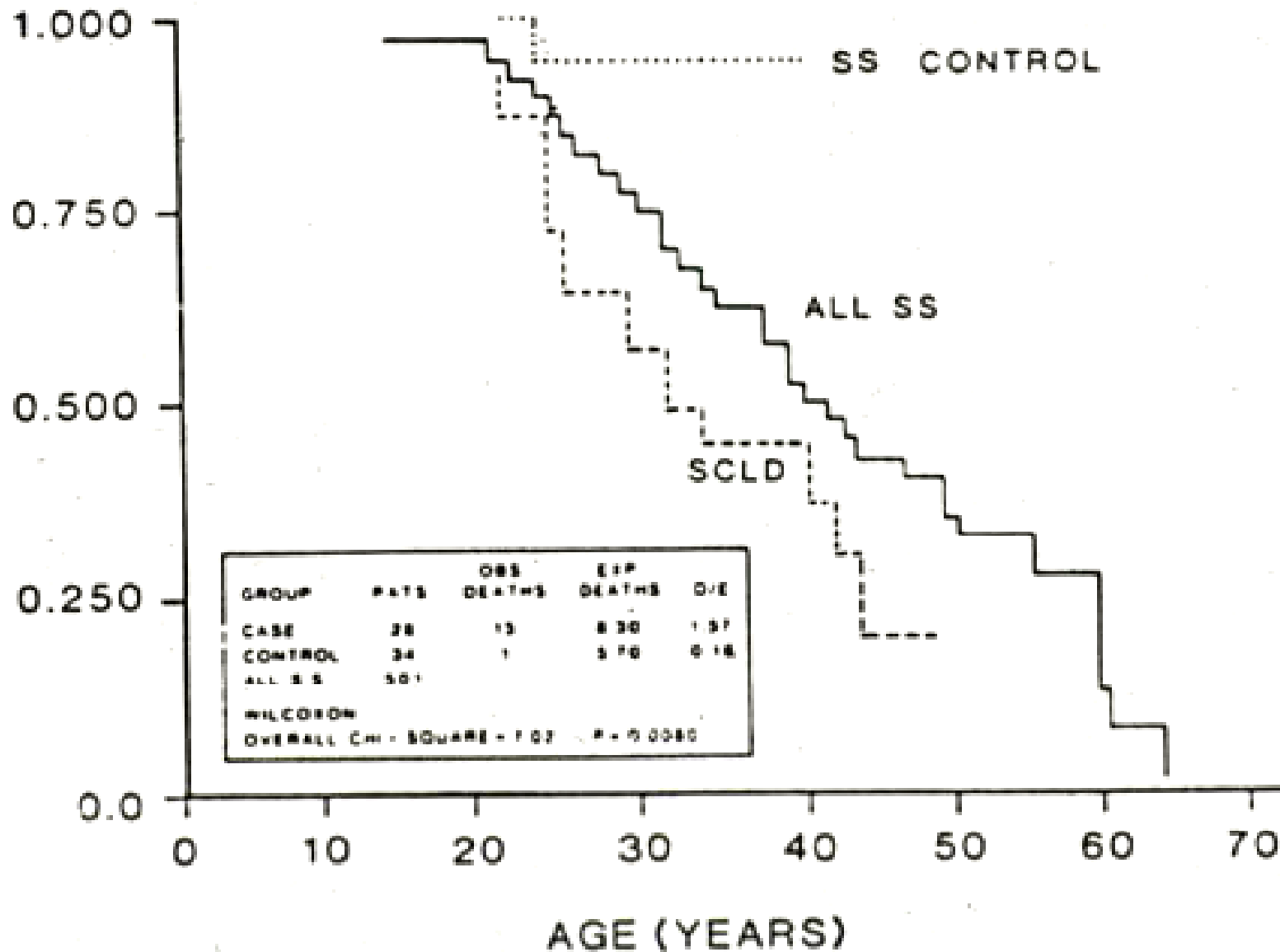


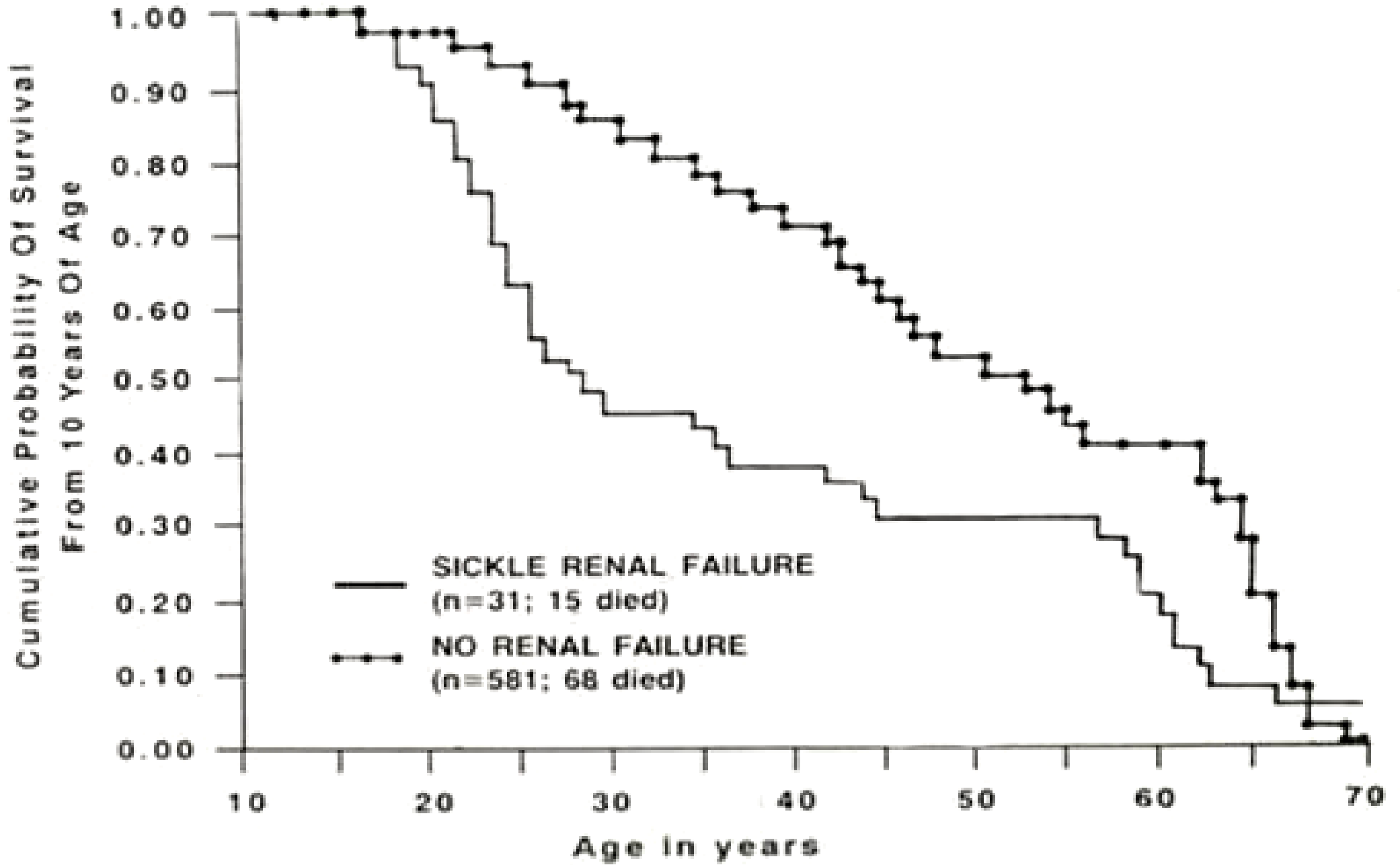


CHRONIC LUNG DISEASE SURVIVAL

COMPARISON OF SURVIVAL FROM AGE 10 TO DEATH (LEFT CENSORED)

PROBABILITY OF SURVIVAL AFTER AGE 10





Outcome of Hb SC

- n = 284; 2,837 patient-yrs since 1964
- Males - 139, Females - 145

Follow-up started at

Age	n	%
< 1 years	43	15
< 10 years	44	15
< 20 years	48	17
> 20 years	149	53

Acute Clinical Events in Hb SC

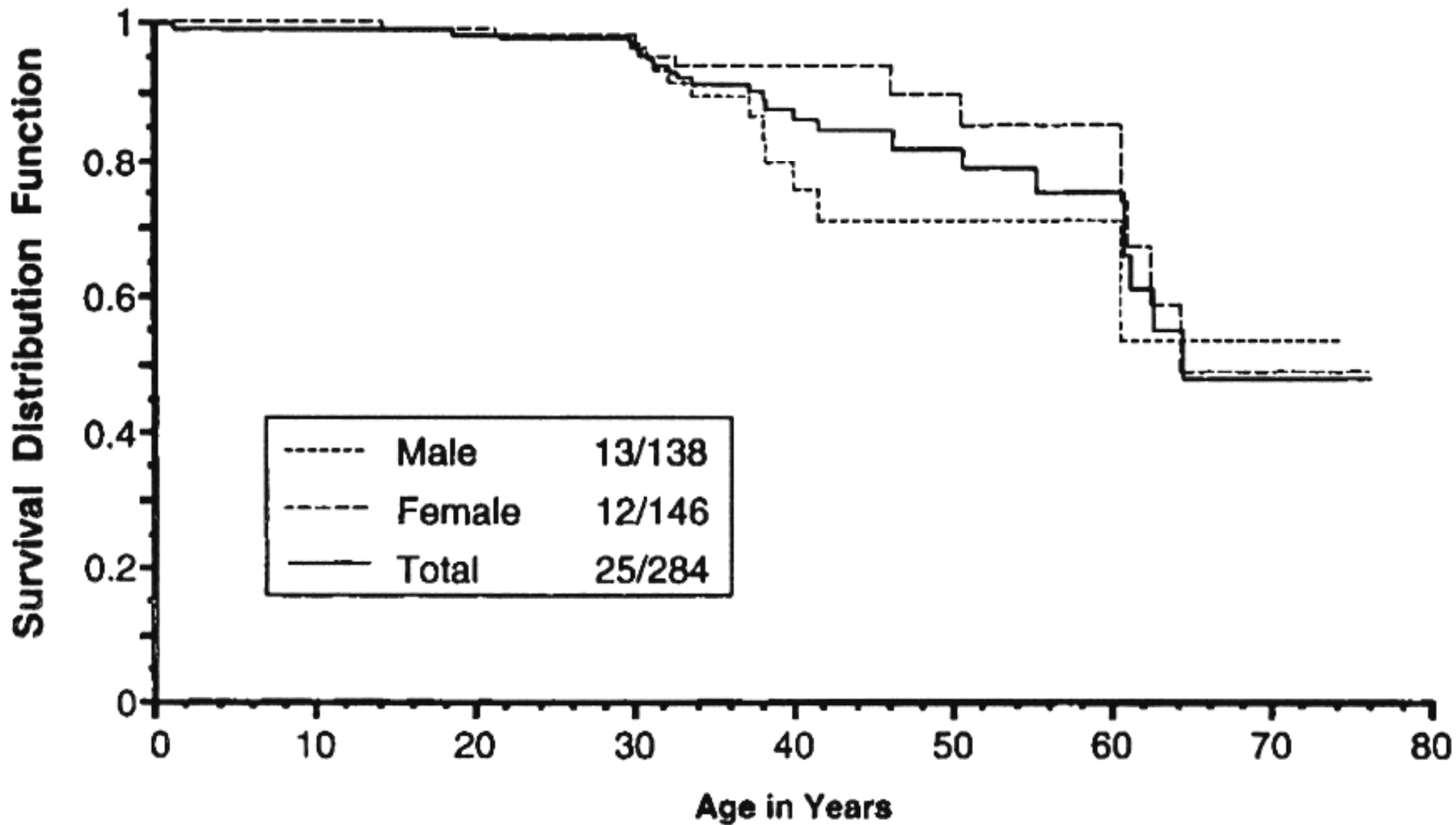
Event	# of pts	% of pts	Incidence per 100 pt-yrs (mean)
Hospitalizations	202	71	47
Sickle related	187	66	38
Uncomplicated pain	157	55	30
2^o sickle pain	95	33	18
ACS	91	32	7
Bone infarct	27	9.5	1.6

Chronic Organ Damage in Hb SC

Organ	# of pts	% pts	Age at Dx (median)
Retina (III, IV)	65	23	28
AVN	42	15	34
Gall bladder	22	8	32
CVA	13	4.6	33
Osteomyelitis	12	4	29
CLD	12	4	32
Renal	8	3	52
Priapism	7	2.5	26
Leg ulcer	6	3	33
Composite	112	39	28
Mortality	25	9	37

Outcome in Hb SC: Mortality

Cause	n=25, 5<20 y of age
Infection	
Hypersplenism	
End-organ failure	5 (20%)
CNS	3 (12%)
Trauma	1 (4%)
ACS/MOF	2 (8%)
Other	12 (48%)
No Dx	2 (4%)



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Options

- Stem Cell Transplantation
- Gene Therapy
- Hydroxyurea
- Transfusion/Chelation

41 y/o woman

- Hb SS Dx age 2
- Cholelithiasis – 1988
- ACS – 1990
- Unable to work since 2004 because of increasing frequency of acute painful episodes
- Referred for evaluation of pain right foot

41 y/o woman

- Hb F 10% on hydroxyurea x ten years
- SpO₂ = 94%, heavy bilateral LL interstitial markings, ↑S2, digital clubbing
- Osteonecrosis T/L vertebrae, both humeral heads



R CR

