Challenges of hemophilia care in a developing country

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Background

Kerala – coastal province in South Western India with population of 35 million, estimated 3500 persons with hemophilia (PwH)

Prior to the opening of the Hemophilia Treatment Centre (HTC), Aluva, Kerala, India in 2014 – PwH travelled 450 km to reach nearest HTC

Lack of factor concentrate availability, affordability

Objectives

To describe the current status, demographics and disease characteristics of hemophilia patients registered at the HTC, Aluva

To describe the relationship of joint status (Hemophilia Joint Health Score, HJHS), independent functional score (Functional Independence Score in Hemophilia, FISH), Quality of Life (QoL)

Methods

Retrospective chart review of data collected from Aluva registry since it's opening in February 2014
Patient demographics, diagnosis and severity of hemophilia
Joint status: Joint bleed data, number and location of affected joints
Outcome measures from validated tools: HJHS, FISH, HemA QoL

Results

Table 1: Number of patients versus type of hemophilia

Type	Number of patients	Percentage (%)
Hemophilia A	433	82.95
Hemophilia B	89	17.05
Total	522	100

Table 2: Hemophilia Severity by Age

| Age category (years) | Hemophilia Severity (Factor Level) | | | Total |
	Severe (<1%)	Moderate (1-5%)	Mild (5-40%)	
<4	22	7	4	33
4-13	62	16	2	80
14-18	45	14	9	68
18-45	158	21	18	197
>45	80	24	23	127
Total	367	82	56	505

Hemophilia Joint Health Score (HJHS) data was available in 127 severe patients.

Average total joint score = 20 ±16.2 (SD). *N.B. higher HJHS = worse joint arthropathy*

HJHS worsened with age (Spearman rho correlation 0.331, p<0.005) (data not shown).

Even at a young age, many patients already had a high HJHS

Highest scoring joints were the knees

FISH score functional domains

self care: 10.9
transfer: 6.4
locomotion: 8.9
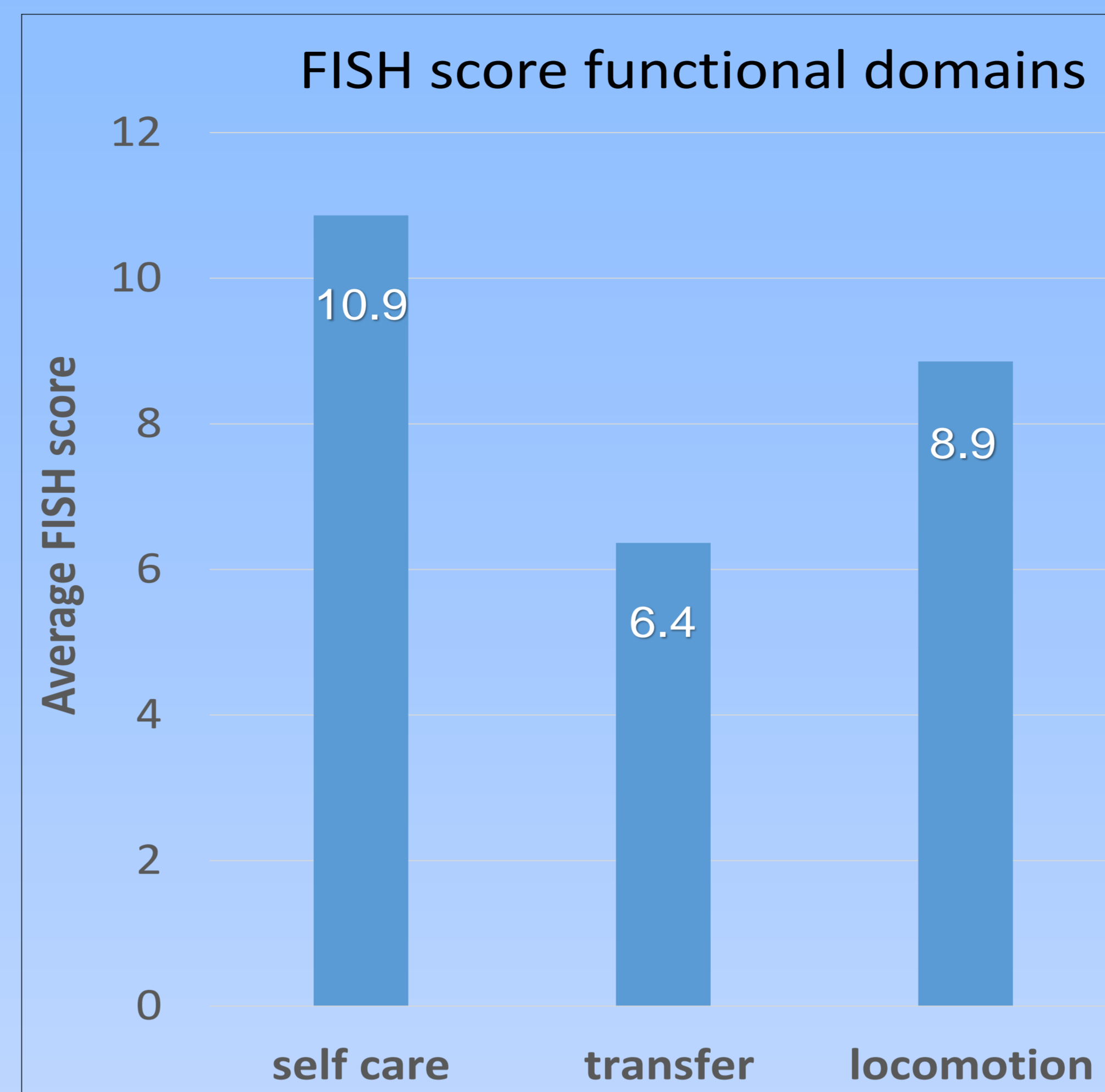
Functional Independence Score (FISH) data was available in 118 severe patients.

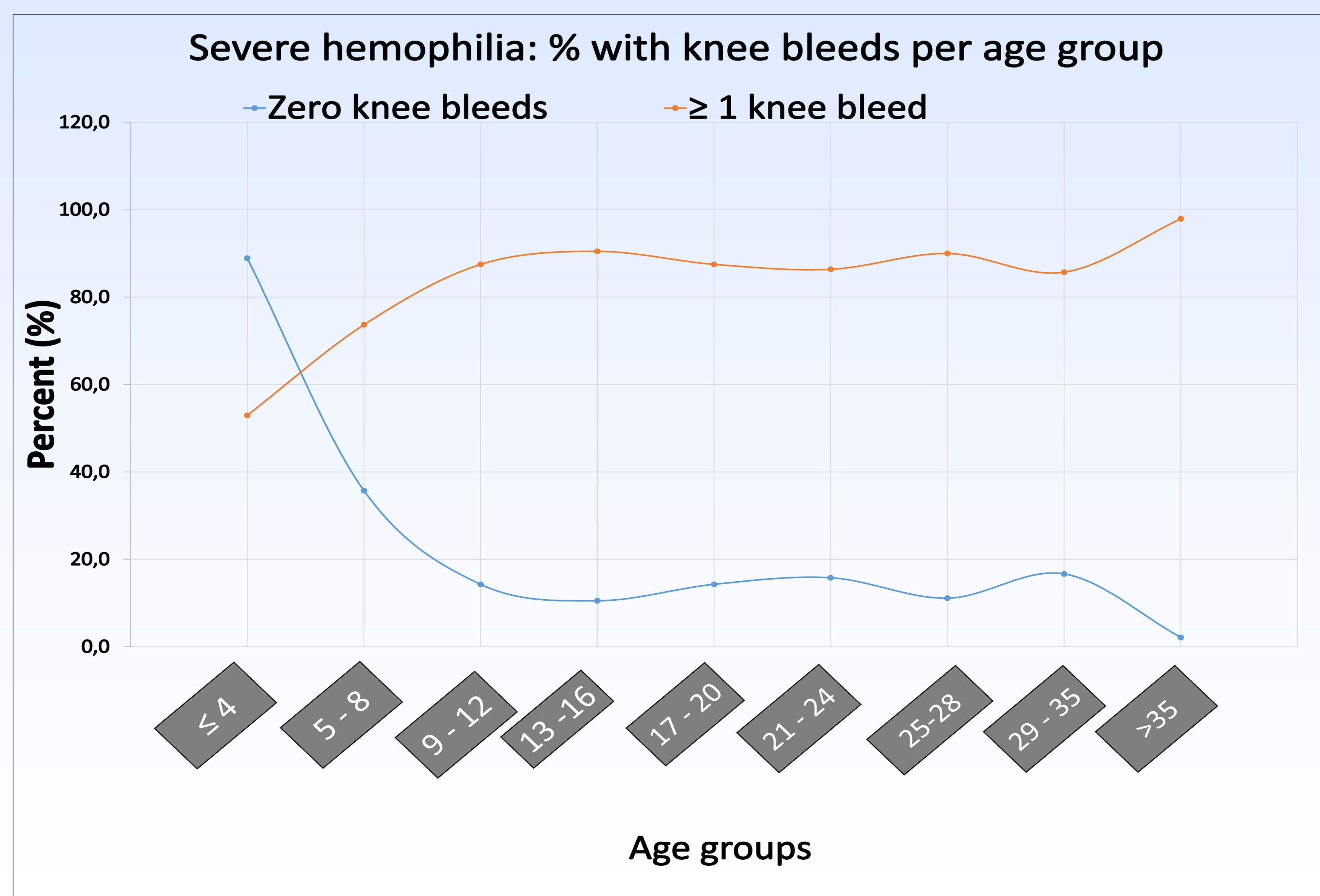Average FISH score 25.9 ±5.9 (SD). *N.B. lower FISH score = worse functional status*

Patients also had worsening functional status with age (data not shown, Spearman rho correlation -0.452, p<0.005)

Lowest functional score was in the transfer domain

FISH score vs HJHS score

Spearmans correlation -0.47(p=0.001)
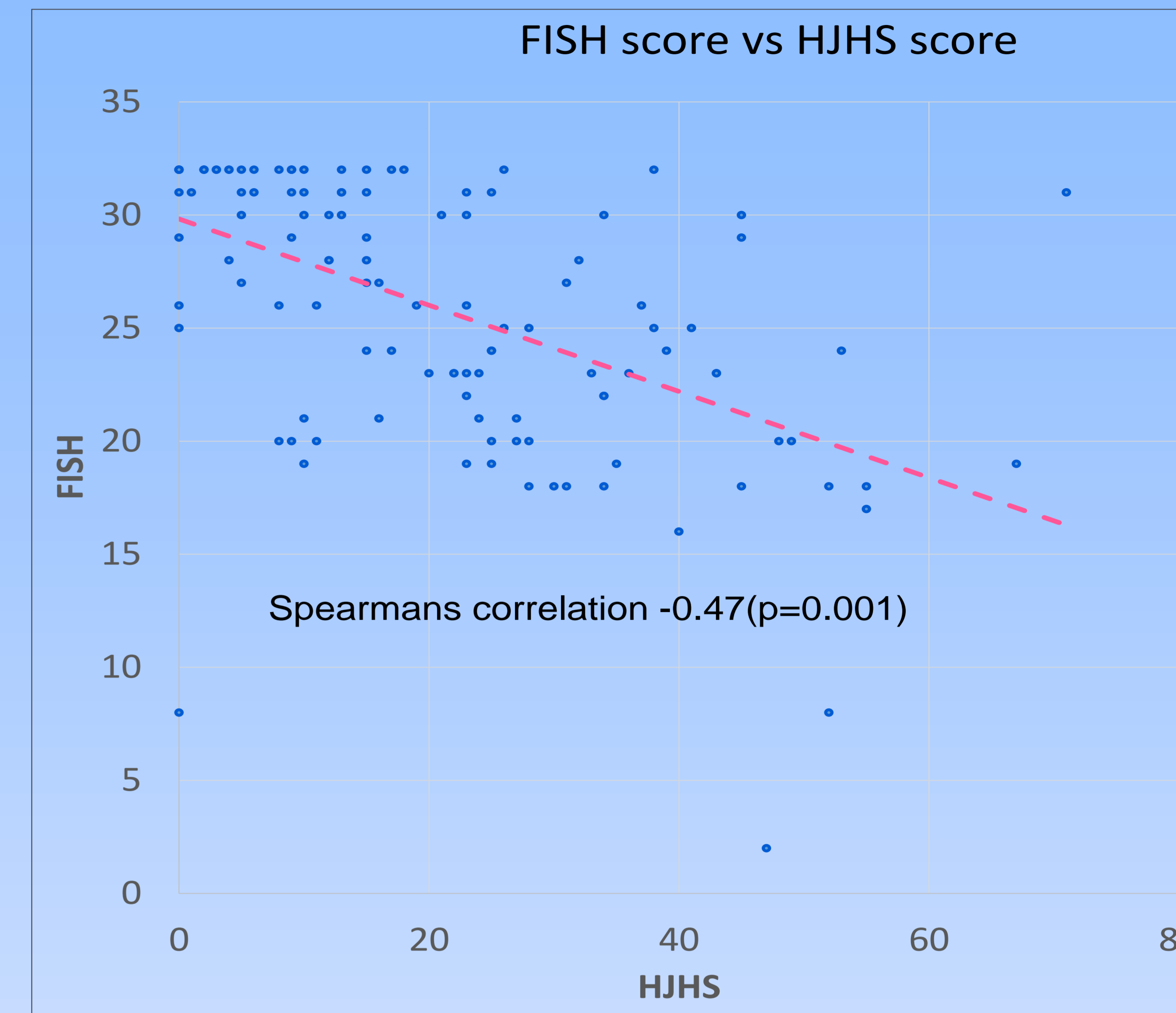
Higher HJHS scores correlated significantly with lower FISH scores in those with severe hemophilia.

This was statistically significant.

Severe hemophilia: % with knee bleeds per age group

Zero knee bleeds — ≥ 1 knee bleed

Age groups: ≤ 4, 5 - 8, 9 - 12, 13 -16, 17 - 20, 21 - 24, 25-28, 29 - 35, >35

By age 9, 85% or more have had a bleed into one or both knees. Data on knee bleeds only, patients may still have bleeds into other joints.

Conclusion

Mostly patients with severe hemophilia are identified in the province of Kerala

Hemophilia patients are young, potentially due to shorter life expectancy or more vigilant diagnosis in children

They have poor joint status starting at a young age

HJHS scores get worse with age

Worse joint status co-relates with worse functional status by FISH

Presumably poor joint and functional status co-relate with poor quality of life but the data is still pending

Future directions

Even low dose prophylaxis will decrease joint bleeding and improve quality of life of PwH.

Funding application submitted to the government to support the low dose prophylaxis study