

Influence of Psychological and Social Factors on Health-related Quality of Life in Carriers of Hemophilia.

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Background

Hemophilia is a severe inherited x-linked bleeding disorder. About 50% of the female carriers have reduced factor levels and are suffering from bleeding episodes themselves. In addition emotional and social problems could have an impact on the Quality of Life (QoL) of carriers.

Patients and Methods

In this multicenter study 318 carriers of Hemophilia A and B with a median age of 27 years were enrolled. 80% were carriers for Hemophilia A, and 70 % of all carriers were affected by the severe form. 78.3% lived in partnerships, 87.7% had at least one child. Carriers had been asked about several issues including bleeding history, family planning, and their social life using an anonymous questionnaire.

Quality of Life (QoL) had been measured by the standardized short version of the WHO-QoL-100-test (bref-test) including the results of a normal female population which served as control group. The test consisted of 26 items and 5 domains including physical, emotional, social, environmental, and global aspects of QoL. A higher amount meant a higher QoL with a maximum of 100.

The study was approved by our local Institutional Review Board (IRB)

Results

The overall QoL was comparable to the normal population. Only the emotional domain showed slightly reduced values (70.24 versus 72.49).

Anxiety occurred in 77% of the carriers and their QoL was reduced (Emotional 69.08 versus 75.34; $p < 0.004$; Social 70.09 versus 79.78; $p < 0.0001$; Global 70.82 versus 76.41; $p < 0.046$) (Fig./Tab.1). **Feeling of guilty** was seen in 47.8% and led in reduced QoL (Emotional 66.93 versus 74.38, Social 68.24 versus 76.49; $p < 0.0001$; Environment 72.00 versus 76.01; $p < 0.013$) (Fig./Tab.2). **Conflicts in the family** (42.5%) were responsible for a reduced QoL (Physical 78.06 versus 82.66; Emotional 66.37 versus 73.79; Social 66.29 versus 77.07; Environment 71.11 versus 75.49; Global 68.14 versus 75.54; $p < 0.008$) (Fig./Tab.3). **Feeling of disadvantages** (Fig./Tab.4) and the **reduced communication about hemophilia** had a negative influence on QoL additionally.

72% carriers were suffering from bleeds like menorrhagia, hematomas, and epistaxis and had a lower QoL (Fig. 5).

	Physical	Emotional	Social	Environment	global
anxiety	77.59	68.68	69.87	71.92	69.85
illness	75.58	65.29	66.19	68.70	64.47
bleeds	79.53	69.33	71.17	72.93	70.83
meno-rhagia	79.12	68.87	69.73	71.81	70.52
hema-toma	78.24	69.31	69.59	72.62	69.63
surgery	79.33	69.67	70.63	72.96	70.90

Fig. 5: QoL in Carriers with or without bleeds. Red: QoL has been reduced significantly in bleeding carriers in comparison to non-bleeding carriers.

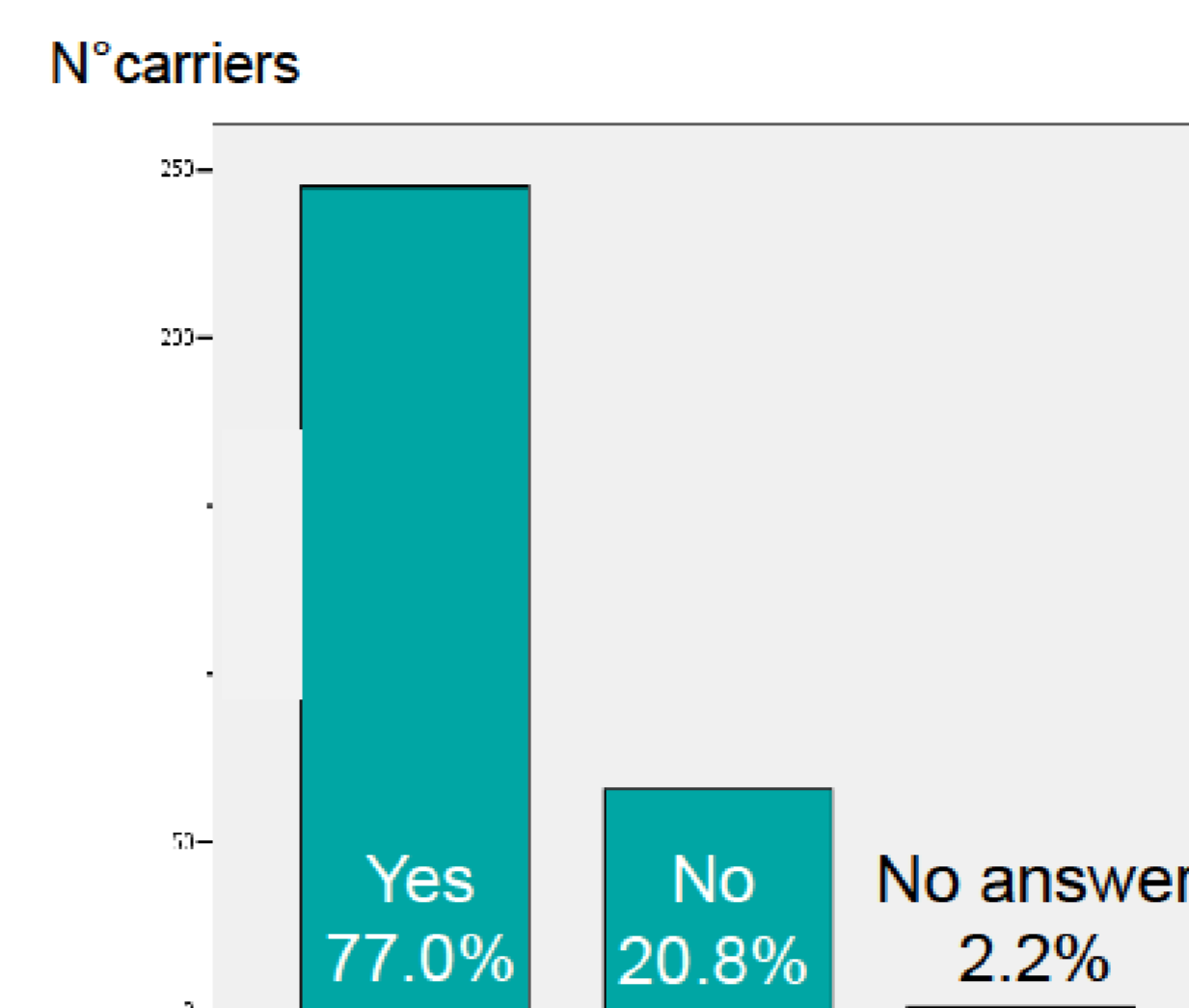


Fig. 1: Carriers feel anxious

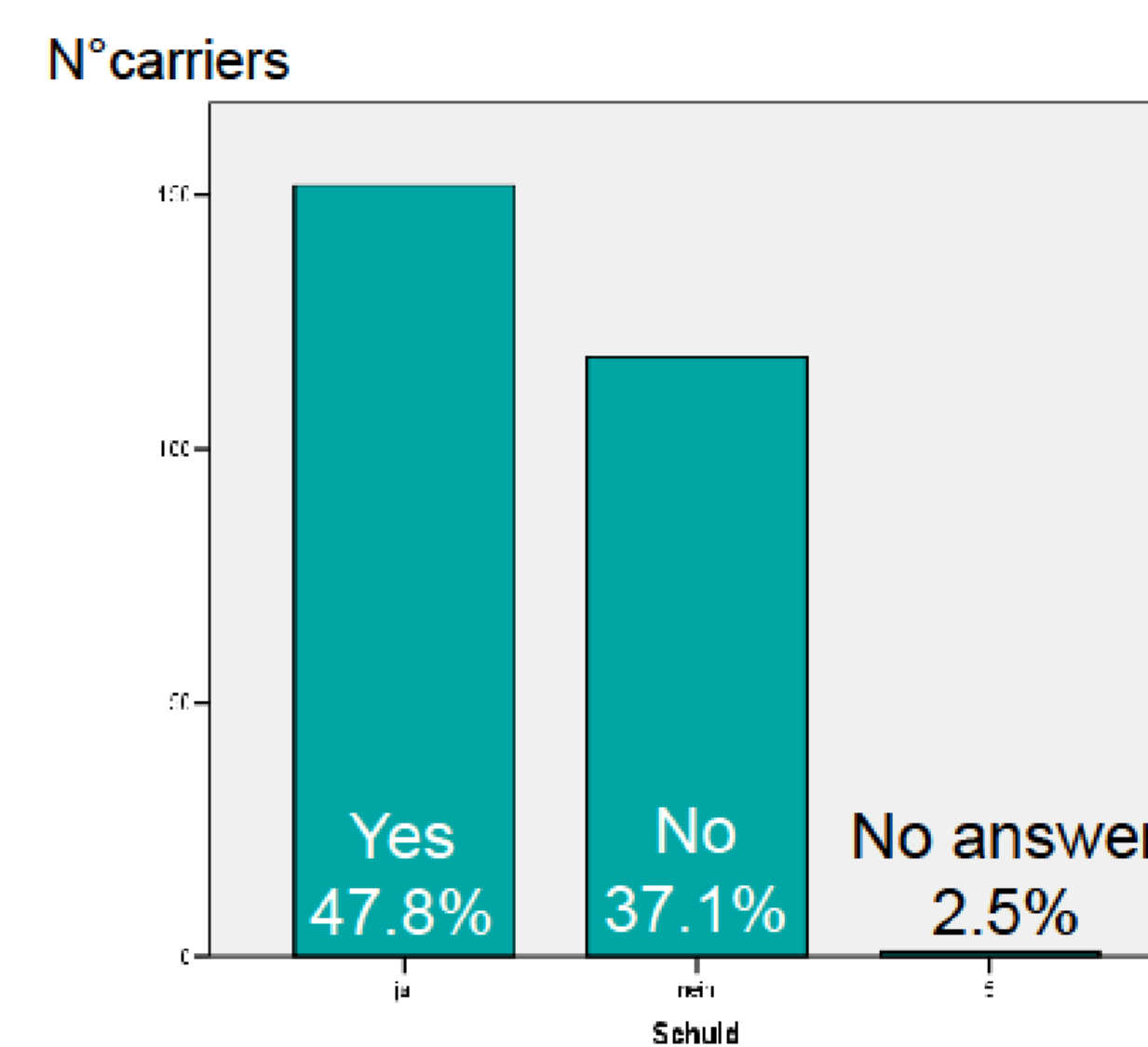


Fig. 2: Carriers feel guilty (12.6% not applicable)

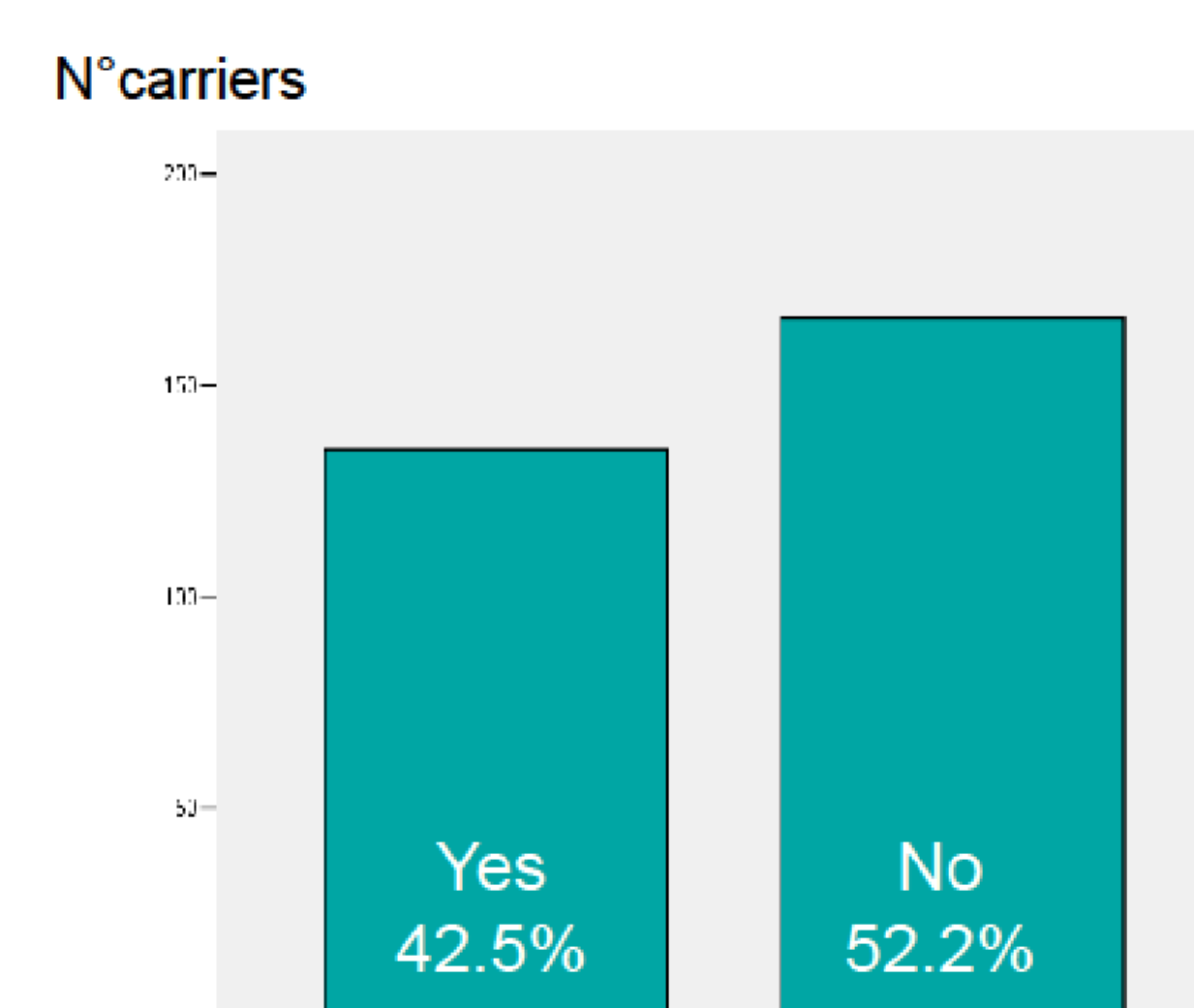


Fig. 3: Carriers having conflicts with their families because their carriership.

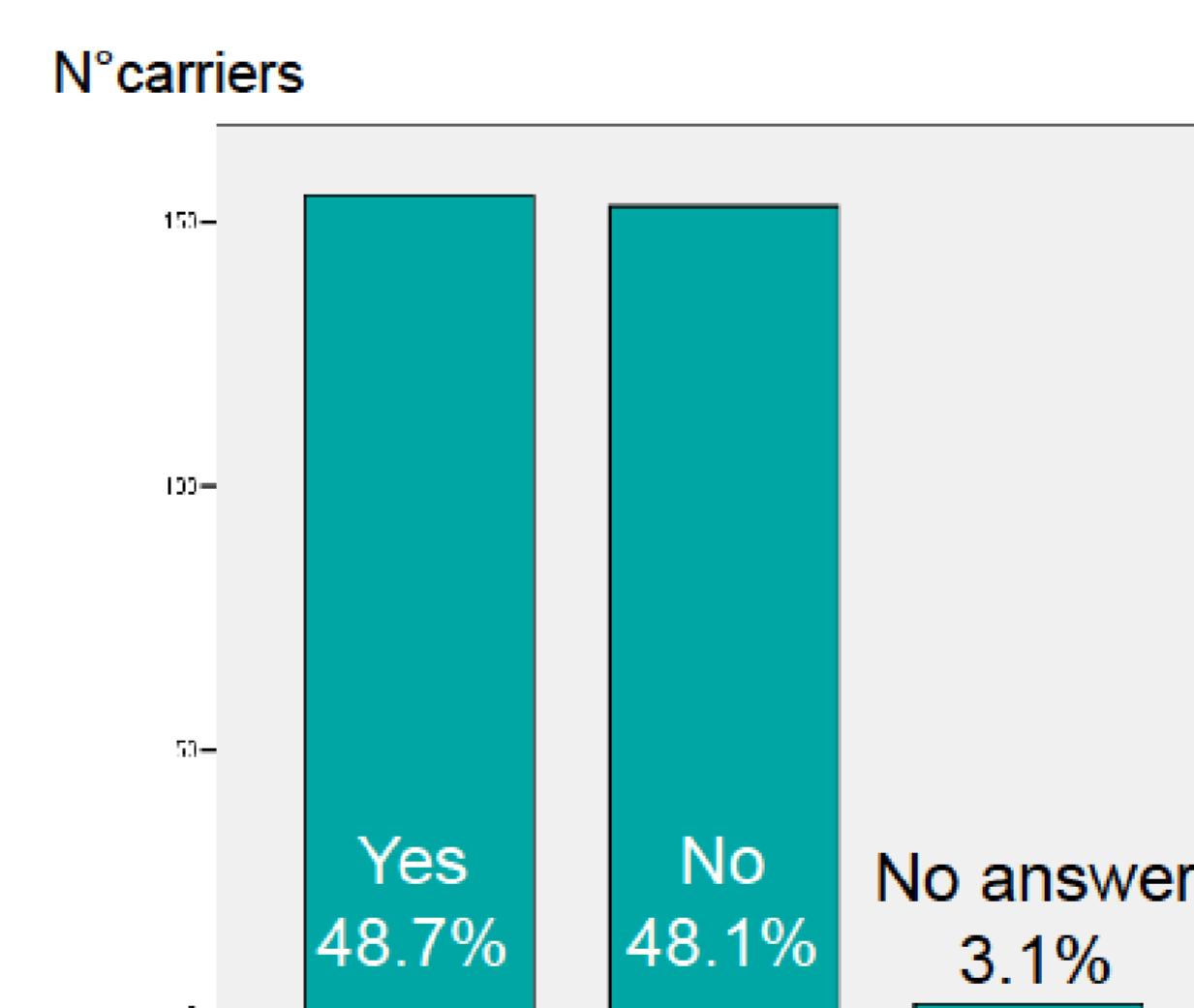


Fig. 4: Carriers feel disadvantaged. Tab.4: QoL of Carriers with (red) or without (black) the feeling to be disadvantaged. $p < 0.0001$, $p < 0.001$

	Physical	Emotional	Social	Environment	global
Mean	79.74	69.08	70.09	72.91	70.82
SD	14.5	14.8	19.1	13.0	19.7
n	244	242	241	241	242
Mean	82.01	75.34	79.78	75.34	76.41
SD	17.8	12.9	12.2	12.8	19.4
N	62	62	61	62	61

Tab.1: QoL of Carriers with (red) or without (black) Anxiety. $p < 0.002$, $p < 0.0001$, $p < 0.046$

	Physisch	Emotional	Social	Environment	Global
Mean	79.63	66.93	68.24	72.00	70.22
SD	14.5	15.8	19.5	13.1	20.7
n	149	149	148	147	149
Mean	80.80	74.38	76.49	76.01	75.00
SD	16.7	12.1	16.4	12.6	18.4
N	116	115	115	115	115

Tab.2: QoL of Carriers with (red) or without (black) the feeling of guilty against their partners, affected sons or carrier daughters. $p < 0.0001$, $p < 0.013$

	Physical	Emotional	Social	Environment	Global
Mean	78.06	66.37	66.29	71.11	68.14
SD	14.8	16.2	19.8	13.3	21.1
n	134	134	133	133	133
Mean	82.66	73.79	77.07	75.49	75.54
SD	14.9	12.4	15.9	12.6	17.68
N	162	162	161	160	162

Tab.3: QoL of Carriers with (red) or without (black) conflicts with their families. $p < 0.008$

	Physical	Emotional	Social	Environment	Global
Mean	77.56	66.33	66.97	70.98	68.10
SD	15.2	15.8	19.3	13.2	19.2
n	154	152	151	152	152
Mean	83.12	74.52	77.60	75.90	75.92
SD	14.6	12.0	14.8	12.3	19.5
N	150	150	149	148	150

Conclusion

Comparing carriers who are worrying about their carriership or the hemophilia in their families, those with feelings of guilty or anxiety, carriers with conflicts in the families or those with problems in their social environment suffer significantly more often from a reduced QoL in contrast to carriers who have not these feelings or problems.

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