

CAUSES OF DEATH IN PATIENTS WITH HEMOPHILIA TREATED AT THE NATIONAL HEMOPHILIA CENTER OF VENEZUELA (1989- 2011)



Apsara Boadas, Mercedes E. Mijares, Luisa Hernández, Arlette Ruiz-Sáez. National Hemophilia Center of Venezuela . Banco Metropolitano de Sangre, Caracas, Venezuela

INTRODUCTION

Since 2001, Venezuela has had treatment for all persons with hemophilia (PWH) with safe clotting factor concentrates (CFC), developing a network of basic Hemophilia Treatment Centers and improving the home treatment program. To evaluate how these advances influenced the mortality of PWH, we performed a retrospective analysis of medical records and compared two periods: 1989 to 2000 and 2001 to 2011.

PATIENTS AND METHODS

We reviewed the medical records of PWH registered at the National Hemophilia Center of Venezuela who died between 1989 and 2011, and analyzed the following factors: age and cause of death, hemophilia type and severity, presence of inhibitor, liver disease (LD) and AIDS. We compared two periods of time: 1989 to 2000 and 2001 to 2011. We calculated the specific mortality rate¹ for patients with hemophilia. Death was categorized according to ICD-10².

RESULTS

In the National Hemophilia Center of Venezuela, 1,546 PWH had been registered, between 1989 and 2000, with a total of 79 deaths in that period of time. The mean age of PWH at the time of the death was 27.4±12.8 (range 2 – 66) years. 5% of the patients had inhibitors. The main cause of death was hemorrhage. Specific mortality rate was 51.09 per 1,000 patients with hemophilia.

During the period 2001 to 2011, we had registered 1,878 PWH, with a total of 60 deaths. The mean age of the patients at the time of the death was 37.2±19.2 (range 1 – 77) years. 8/60 (13%) patients had inhibitors and the main causes of death were intracranial hemorrhage, violent acts (homicide, car and motorcycle accidents), or drug abuse. Specific mortality rate was 31.94 per 1,000 patients with hemophilia.

Table 1. Type and severity of the PWH who died between 1989 - 2011.

Type of hemophilia	1989 to 2000	2001 to 2011
Mild HA	2	7
Moderate HA	42	21
Severe HA	32	18
Mild HB	0	7
Moderate HB	3	3
Severe HB	0	4
Total	79	60

Figure 1. Type and severity of hemophilia comparing 1989 – 2000 and 2001 – 2011.

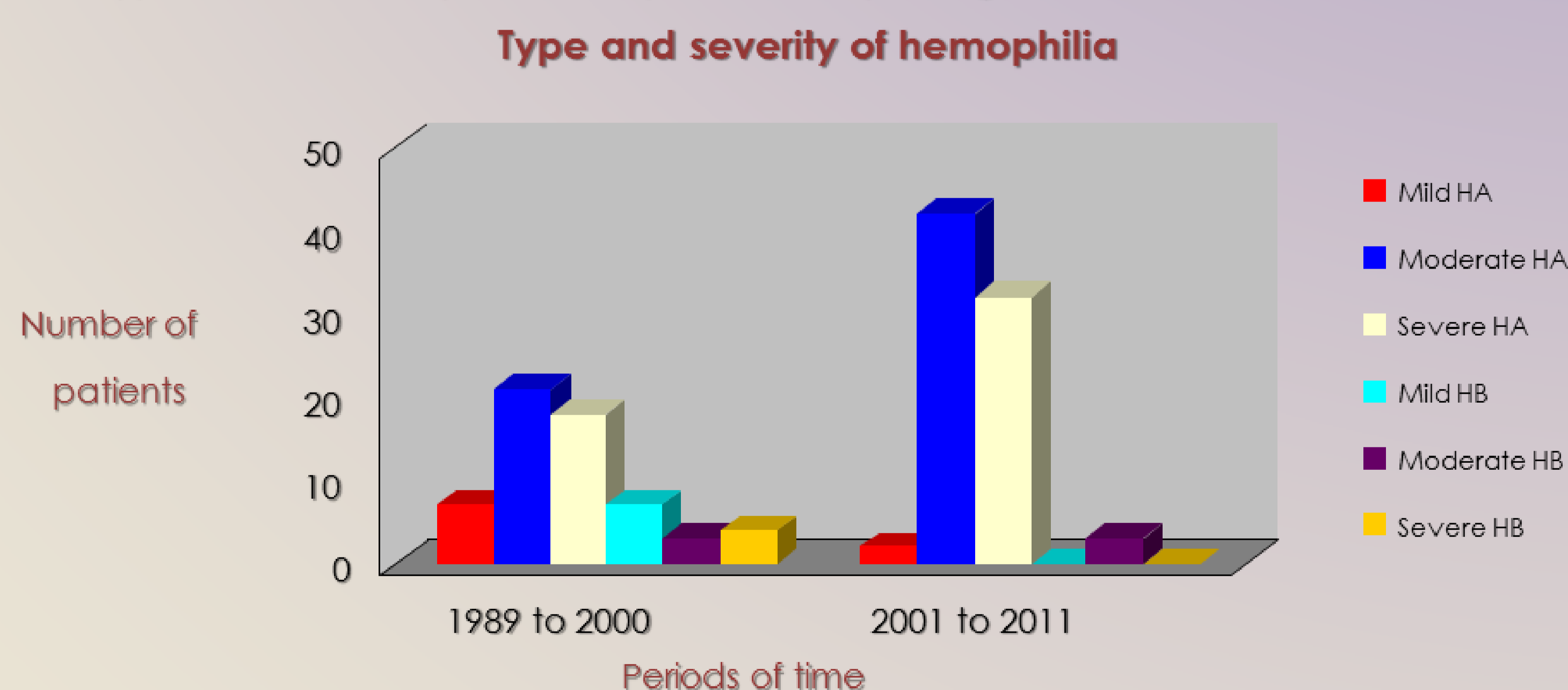


Table 2. Age at time of death of the PWH comparing 1989 - 2000 and 2001 - 2011.

Age at time of death (years)	1989 to 2000	2001 to 2011
0 to 2	4	1
3 to 10	4	2
11 to 17	16	4
18 to 25	16	14
25 to 32	15	10
33 to 40	8	6
41 to 48	8	9
49 to 56	3	4
57 to 64	3	5
65 to 72	2	2
>73		3

Figure 2. Age at time of death of the PWH between 1989 and 2011.

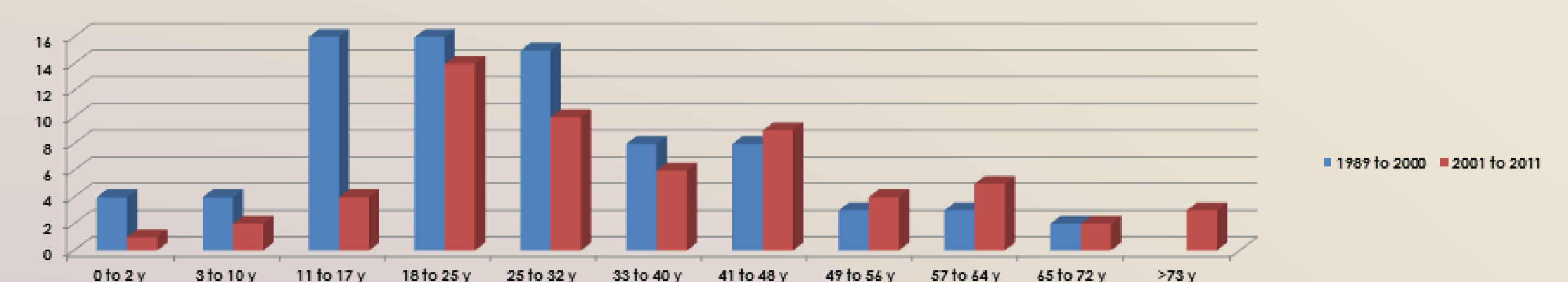


Table 3. Causes of deaths comparing the periods 1989 - 2000 and 2001 - 2011.

CAUSE OF DEATH	1989 - 2000	2001 - 2011
	n(%)	n(%)
CARDIOVASCULAR	2 (2.5)	3(5)
HEMORRHAGE	No Intracranial hemorr.	4(6.7)
	Intracranial hemorr.	16(26.7)
INFECTION	SEPSIS	2 (2.5)
	AIDS	40 (50.6)
	HEPATITIS RELATED	8 (10.1)
		7(11.7)
NEOPLASM	0	3(5)
VIOLENT	Homicide	5 (6.3)
	Car or motorcycle accident	1(1.3)
	Drug abuse	2(2.5)
UNKNOWN	2(2.5)	0
TOTAL	79(100)	60(100)

Figure 3. Causes of death comparing the periods 1989 - 2000 and 2001 - 2011.

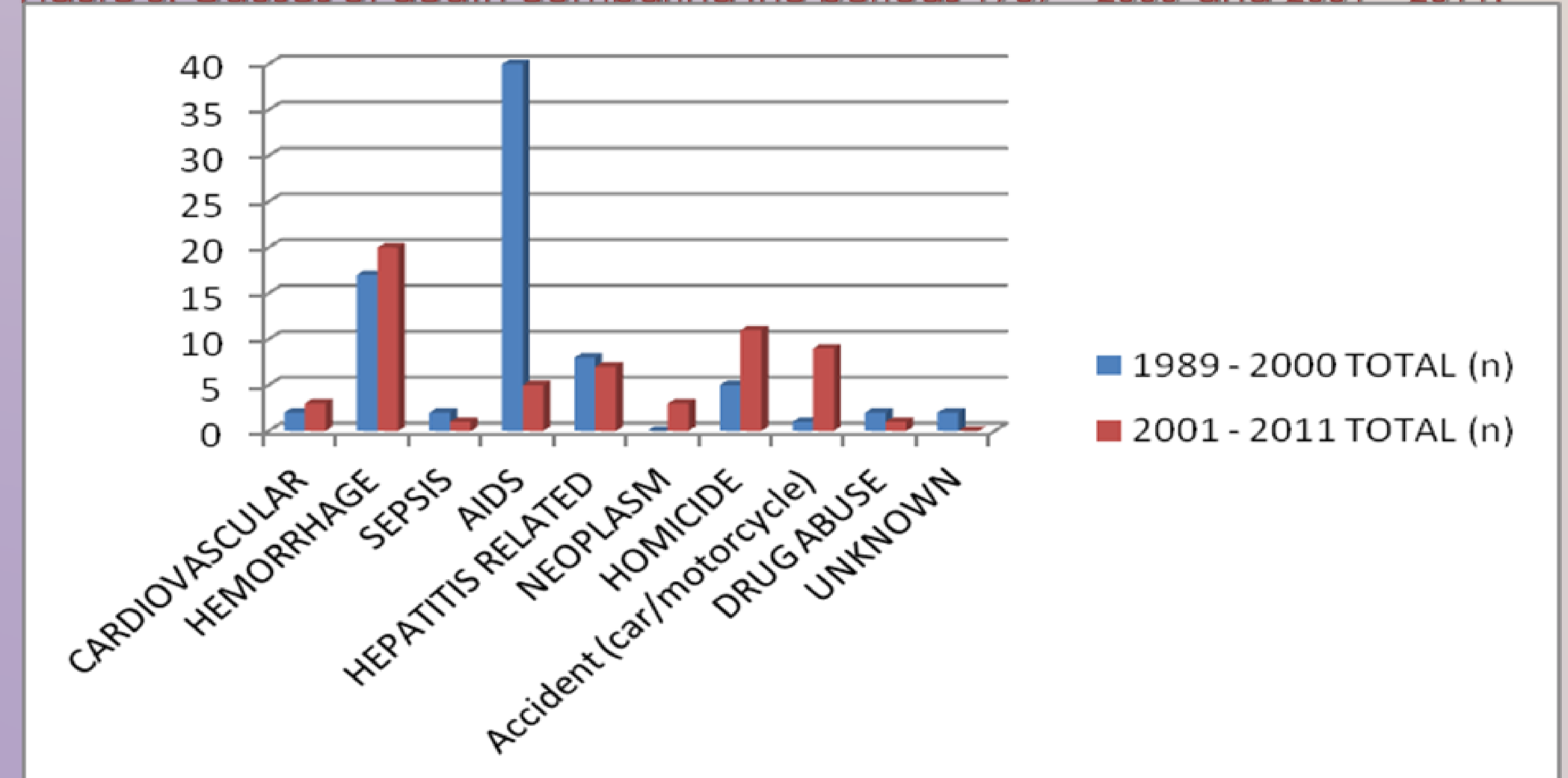
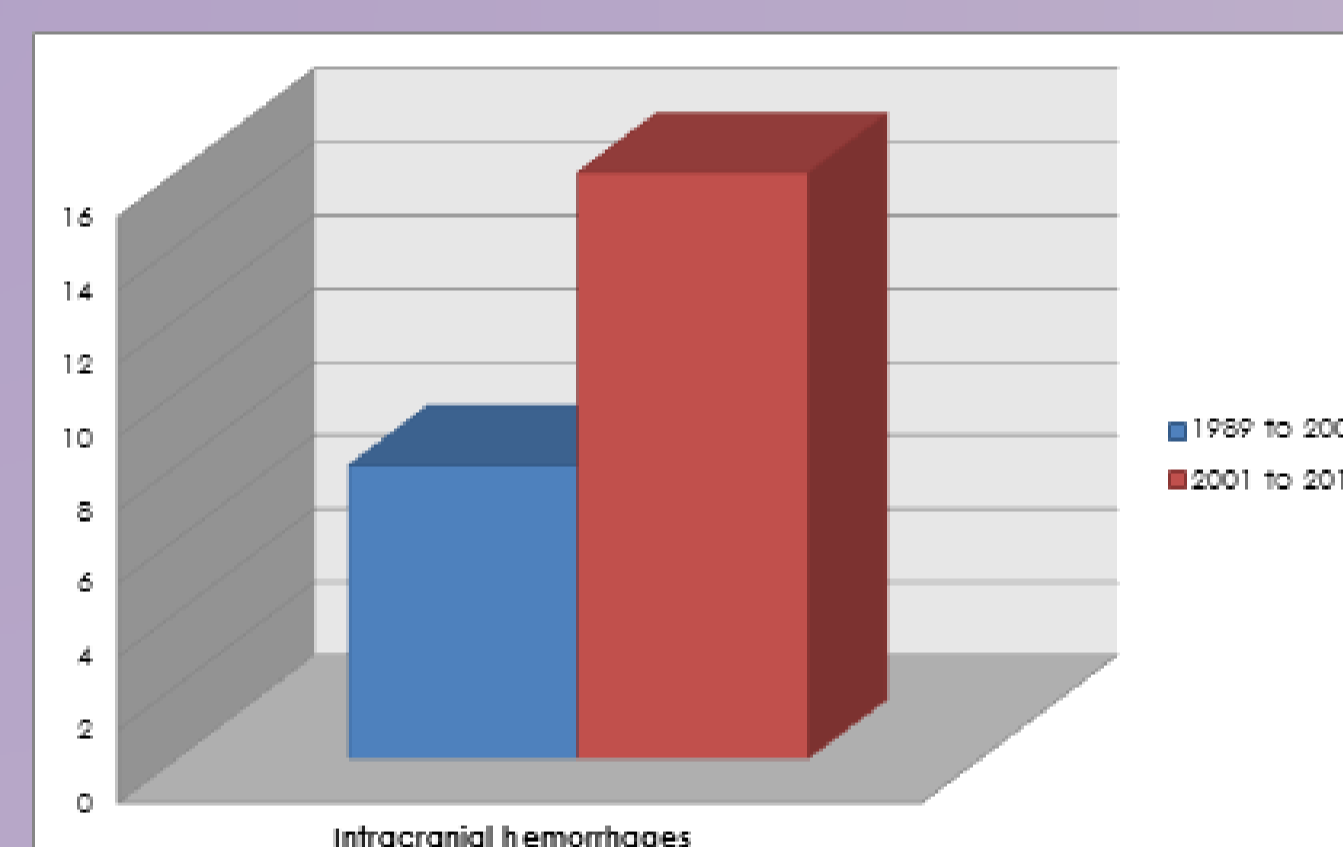


Figure 4. Deaths by Intracranial hemorrhages comparing 1989 – 2000 and 2001 – 2011.



DISCUSSION

Mortality due to liver disease was similar in both periods of time (1989 – 2000 and 2001 – 2011) as would be expected, because of the high rates of hepatitis C in the person with hemophilia during 1989 to 2011. From 2001 to 2011 we also observed a decline on AIDS-related deaths and the age at the moment of the death was 10 years higher ($p < 0.0075$), after the use of safe CFC, while liver disease-related deaths remained constant.

The occurrence of intracranial hemorrhage and especially violent deaths was higher during 2001 – 2011 than the previous decade, and it is a consequence of the safety issue in our country. Death as a result of hemorrhages due to violence (homicide or car and motorcycle accidents) in the hemophilia population, were similar to those reported at the general population in Venezuela.

The educational programs of home therapy with CFC established and promoted by the National Hemophilia Center may also have influenced in mortality reduction because it facilitates early treatment of bleeding episodes.

CONCLUSIONS

- It is necessary to consolidate campaigns to avoid dangerous behavior in PWH.
- Number of death in mild hemophilia patients could reflect an increase in registered cases.

References

- [Crude death rate \(per 1,000 population\)](#) based on *World Population Prospects The 2008 Revision*, United Nations. Retrieved 22 June 2010
- World Health Organization. Manual of the international statistical classification of diseases, injuries, and causes of death, ninth revision. Geneva, Switzerland: WHO; 1977.

