

Current diagnosis status of children in China: An analysis of single center data including 417 cases of children with hemophilia in 13 years

Authors: Ling TANG, Runhui WU*, Xinyi WU, Yingzi ZHENG, Zhenping CHEN, Gang LI, Yan WANG, Ningning ZHANG, Jishui ZHANG, Yun PENG

Hospital: Hemophilia Work Group, Beijing Children's Hospital affiliated to Capital Medical University, Beijing, China

Correspondence:

Dr. Runhui WU, Hemophilia work-group, Hematology Centre, Beijing Children's Hospital affiliated to Capital Medical University, No.56 Nanlishi Road, West district, Beijing 100045, China.
Tel: +86 10-59612600 Fax: +86 10-59718700 e-mail: runhuiwu@hotmail.com

OBJECTIVES

In China, with poor understanding of hemophilia, many patients can not be diagnosed in early stage. This delay leads to postponing of treatment. With the foundation of HTCCNC (Hemophilia Treatment Center Collaboration Network of China, the diagnostic situation have been improved through years. However, there is still a gap of early diagnostic ability between China and developing countries

Hemophilia cases from our treatment center since 2007 were retrospectively analyzed about the information of onset and diagnosis, to comprehend progress of Chinese hemophilia care and children hemophilia diagnosis present condition in our country, and provide guidance for hemophilia treatment scheme which accords with the situation of China.

METHODS

Collecting hemophilia children cases in our hospital outpatient from January 1, 2007 to May 30, 2013, to discuss the diagnosis time and its influence factor.

Severity of first hemorrhage: mild: like petechia that do not need treatment or diagnosed through screening test before operation; moderate: joint/muscle bleed, hemorrhage in operation, mucosa bleeding or hematoma limiting joint/muscle function; severe: bleed threaten to life, like intracranial bleeding or hemorrhagic shock

Diagnostic timing (interval between first bleed and diagnosis): short-interval: ≤ 1 month; mediate-interval: 1-6month; long-interval: >6 month

Area level: I: area where HTCCNC 6 main centers locates (Tianjin, Guangzhou, Shanghai, Beijing, Hefei, Jinan); II: cities besides these 6 cities; III: rural area

RESULTS

Data of 417 cases were collected, respectively from 24 provinces or autonomous regions; Median age was 2.5 years (0.1 to 13.1); Hemophilia A were 333 cases (79.9%), hemophilia B were 84 (20.1%); 33 were mild (7.9%), 204 were moderate (48.9%), 180 were severe (43.2%) 104 children had hemophilia family history (24.9%)

First bleeding situation: occurred at the median of 10.0 months (0.0-89.0 months); Mild hemorrhage in 181 cases (43.4%), moderate in 197 cases (47.2%), severe in 39 cases (9.4%)

Reason of first bleed: 27 (6.5%) was iatrogenic hemorrhage, 12 (44.4%, 12/27) were diagnosed through screening test before operation, 3 (11.1%, 3/27) blood during parturition

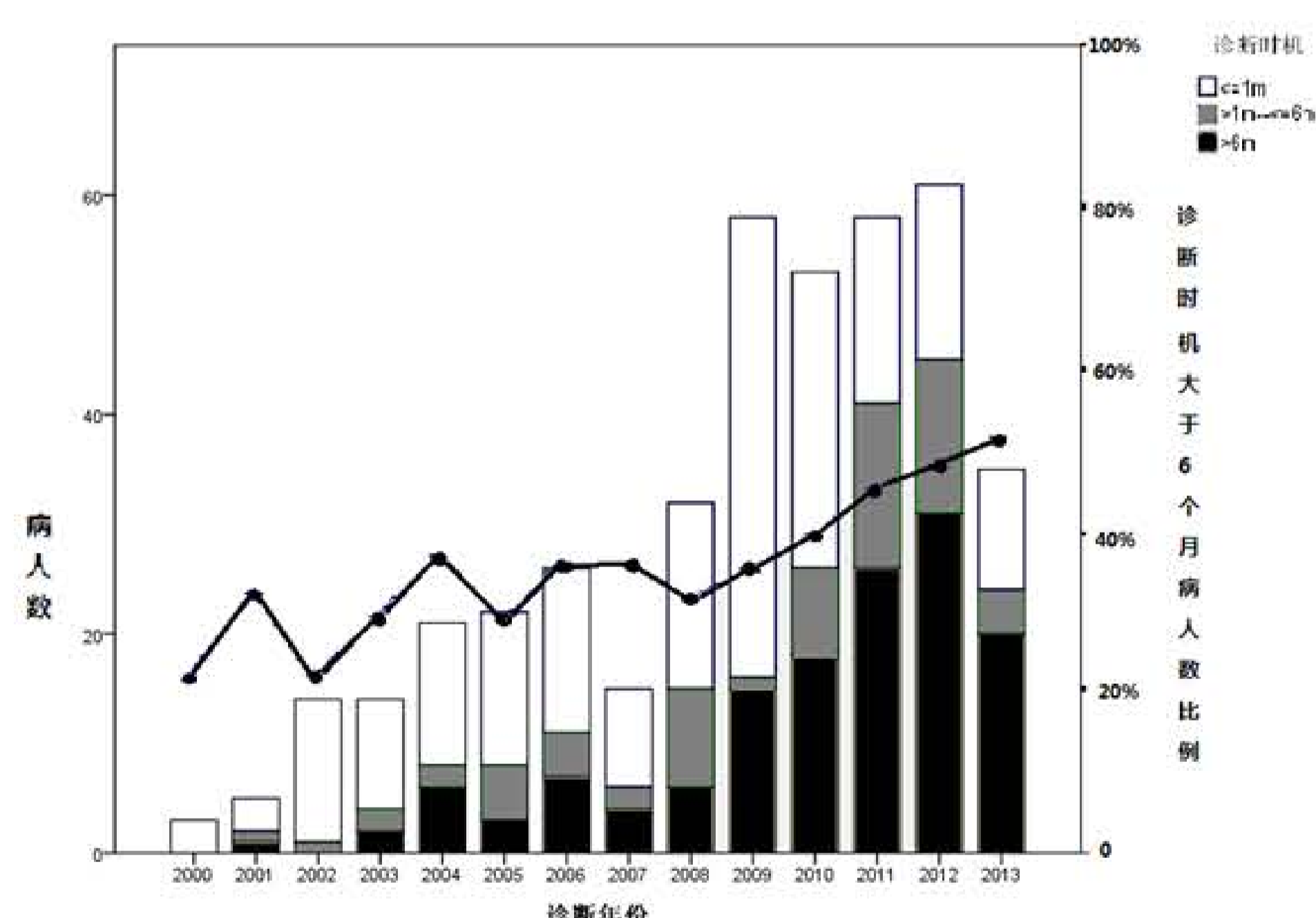
Average diagnosed in 22.5 ± 14.8 months after birth (0.0-178.0 months); Average interval between first bleed and diagnosis was 9.10 ± 0.82 months (0.0-100.0 months)

Diagnostic timing: 210 cases (50.4%) of short-interval, 68 (16.3%) of mediate-interval, and 139 (33.3%) of long-interval. Diagnostic timing had no difference between children with and without family history ($P=0.71$), neither between children from different level of area ($P=0.281$); Patients with more severe bleed had higher rate of short-interval diagnostic timing ($P=0.012$). No difference between children with moderate and severe first bleeding ($P=0.463$).

In recent 12 years, the number of new diagnosed cases has a trend of increase. The percentage of short, mediate and long interval had difference in recent 3 years ($X^2=14.1, P=0.007$); the long-interval percentage increased in recent 3 years, they were 44.8%, 50.8% and 57.1% respectively.

Graphs and tables

Year	2000	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012	2013
number of diagnosed	3	5	14	14	21	22	26	15	32	58	53	58	61	35
III area%	66.7	40.0	42.9	28.6	42.9	45.5	46.1	53.3	50.0	48.3	54.7	50.0	59.0	57.1
Diagnosed interval														
short%	100.0	60.0	92.9	71.4	61.9	63.6	57.7	60.0	53.1	72.4	50.9	29.3	26.2	31.4
mediate%	0	20.0	7.1	14.3	9.5	22.7	15.4	13.3	28.1	1.7	15.1	25.9	23.0	11.4
long%	0	20.0	0	14.3	28.6	13.6	26.9	26.7	18.8	25.9	34.0	44.8	50.8	57.1
III area and long interval%	0	0	0	0	2.6	1.3	2.6	2.6	6.6	11.8	9.2	19.7	23.7	17.1



CONCLUSIONS

There is obvious progress of haemophilia care in China. The number of diagnosed haemophilia children increased rapidly in recent years; patients with more severe first bleeding are prone to have earlier diagnosis. However delay of diagnosis is still ubiquitous. We need more publicity for haemophilia and disseminate knowledge of this rare disease, especially in less developed areas, in order to improve people's understanding of the disease and ability of early diagnosis

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