

# The needs and support measures of Japanese haemophilic carriers : literature review and discussion

Tomie Fujii<sup>1)</sup>, Mayuko Ishikawa<sup>2)</sup>, Toshio Kobayashi<sup>1)</sup>, Teruhisa Fujii<sup>3)</sup>

1) Graduate School of Biomedical & Health Sciences, Division of Nursing Science, Hiroshima University, Japan

2) Oki Health Center, Oki branch office, Shimane Prefecture, Japan

3) Division of Blood Transfusion, Hiroshima University Hospital, Japan

## Introduction:

Haemophilia is an X-linked recessive inheritary disease passed from carriers to offspring. Therefore, some female families of haemophiliacs may have much anxiety at their pregnancy and delivery. Although haemophilia patients came to be able to do healthy life because of the medical development, there are few reports in which medical care providers focus on haemophilic carriers (HCs) in Japan. The Japanese haemophilia comprehensive medical care system is very immature in comparison with that in American and European countries. We reviewed and analysed literatures on the support of HC in Japan to clarify the support measures needed for them.

## Methods:

The search was intended for 1995 through 2011. The literatures were investigated with a search engine for medicine and nursing using the key words 'haemophilia', 'carrier', or 'nursing', and 10 papers were selected as the subjects of our study. Carriers' needs and support situations for them were sampled and compared with those in studies from developed countries in the field of haemophilia.

## Results:

Table 1 The comprehensive care system of hemophilia in Japan

| Methods   | Results   | References           |
|---|---|----------------------|
| Subject : medical care providers, hemophilic patients and the family<br>Method : a questionnaire survey | The facilities in which were examined less than ten patients were 24 of 52.<br>There were 19 of 52 facilities established the special outpatient clinic for hemophiliacs. The facilities which locate comprehensive care for outpatients were 5 of 52.<br>Out of 24 facilities which established the comprehensive care system of hemophilia, 3 were engaged in registered nurses and none was engaged in special nurses. | O. Ono, et.al. 1999  |
| Subject : hemophilic patients and the family<br>Method : a questionnaire survey                         | There were 5 facilities which provide the comprehensive care for hemophiliacs.<br>There was only one facility which assigned coordinator nurse.   | O. Ono, et.al. ,2003 |
|   | For establishment of the comprehensive care system. Promotion of nurse specialized in haemophilia<br>Departments which should be cooperated with<br>-orthopedics -brace companies<br>-rehabilitation -dentistry<br>-which provide the opportunity of medical treatment to medical residents<br>-Others(Pharmacists, clinical psychologists, Radiology, Social workers, Otolaryngology, et.al)                             | T. Oka.2005          |

Facilities experienced less than 10 patients were accounted for half, and the facilities placed a specialized outpatient clinic and assigned nurses were a few.

Table 2 The needs and support measures of HCs in Japan

| Methods   | Results   | References              |
|---|---|-------------------------|
| Subject : hemophilic patients and their families<br>Method : a questionnaire survey   | By the question whether you think to want to consult with a nurse about "heredity, pregnancy and delivery"...<br>Persons with experience like that: 15/142 (10.6%) .<br>Persons without experience: 47/248 (19.0%)<br><br>By the question about annoyance due to haemophilia, -Any person who did not provide knowledge and information of the illness : 18/393 (4.6%).   | O. Ono, et.al. 2003     |
| Subject : Mothers of hemophilic children<br>They were divided into 12 definitive haemophilic carriers and 12 possible haemophilic carriers<br>Method : a questionnaire survey | A survey for 24 persons(12 definite carriers and 12 possible carriers)<br><b>Notification time and a person of notification:</b><br>Among 3 HCs with family history which their fathers were hemophiliacs, there was a person notified before the delivery and were two announced after it.<br><b>Genetic counseling:</b><br>Twenty-one of 24 persons were explained in detail about the heredity from doctors. On the time of explanation, most persons answered "when a child diagnosed as haemophilia".<br><b>Diagnosis of carrier:</b><br>The persons with experience examined such as coagulation factor activity levels were 7 of 12 definite carriers, and 9 of 11 possible carriers.<br><b>Prenatal diagnosis:</b><br>One of 24 received a prenatal diagnosis.<br>Six persons hoped if possible in reply,<br>It was a reason when "you should know the risk at the time of the delivery beforehand" | Y. Matsuo, et.al. 2011. |

Haemophilic patients and their families desired knowledge of "heredity, pregnancy and delivery". It was discovered that a half of the HCs in the studies had not known that they were carriers until their deliveries.

Table 3 Genetic counseling for HC in Japan

|             | Results  | Reference               |
|-------------|--|-------------------------|
| Literature1 | A HC who was pregnant hoped a prenatal diagnosis of the baby. If it was a boy, she wanted to have an abortion. It was judged that this case was inapplicable to the prenatal diagnosis. The reasons why were that a haemophilic was possible to be a social life because of the development of medical care and a social disadvantage for them had improved having good there being an effective cure.     | S. Nakagomi et.al.,2003 |
| Literature2 | A fiance with HC had a considerably negative preconception toward hemophilia. He was accepted by twice of counseling. Some clients may have excessive anxiety because they cannot judge appropriate information about inherited diseases by the information obtained from the internet.  | T. Shinke 2005.         |
| Literature3 | There were three HCs whose fathers had haemophilia. Explanation about The possibility of being born a hemophilia child and the delivery risk was explained and an applicant for a genetic counseling was counseled. One of them became the vaginal delivery by the premature delivery, and two of them were delivered by a Caesarean section. There were none of the babies who had intracranial bleeding. | Y. Hattori, et.al.,2010 |

The persons who needed genetic counseling in HCs used genetic medical care.

## Discussion:

The fact that few institutions can provide special care for haemophiliacs suggests that opportunities for counselling patients about their unique problems, even though they have troubles or anxieties, are limited. Patients' and the family's needs in terms of genetics, pregnancy, and delivery appear to be in the department of genetic medicine at these institutions. The viewpoint that HC should be cared for as potential haemophiliacs in Japan is lacking. It is important to spread knowledge of HC to the fields of medicine and nursing to improve the present situation. In future, we will perform the investigation about the support system which HCs need themselves.

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