Original Article
ISSN (Online): 2582-7472

ShodhKosh: Journal of Visual and Performing Arts
June 2024 5(6), 1621–1625

COMBATING HAEMOPHILIA: SOME LESSONS FOR INDIA

Mrs. Shabnam Mushtaq [1], Dr. Wakar Amin Zargar [2]

[1] Ph.D. Candidate, Department of Social Work, University of Kashmir, Naseem Bagh Campus, Hazratbal Srinagar Jammu and Kashmir, India
[2] Assistant Professor, Department of Social Work, University of Kashmir, Naseem Bagh Campus, Hazratbal Srinagar Jammu and Kashmir, India

ABSTRACT

Haemophilia is a sex a linked heredity bleeding disorder that predominantly affects male population and while women are carriers of the affected gene. Approximately 4 lac people worldwide are affected with this disorder. The response to this disease involves a range of treatment interventions involving proper treatment protocol, physiotherapy and comprehensive rehabilitation programmes. However, budgetary constraints, geographical and socio-cultural factors overburden the process of delivering adequate treatment for hemophilia patients in different countries the socio-economic inequalities among the countries have been found to greatly influence the treatment, management and care of heamophilia patients. Thus, post industrialized nations have enough accessibility to various treatment resources, while as the emergent countries like India have excelled in the field of physiotherapy and rehabilitation programs. The authors are of the view that for optimal heamophilia treatment care and management, a comprehensive strategy involving multiple interventions is required. This paper aims to suggest a brief description of strategies required to achieve an optimal heamophilia care in India.

Keywords: Disease, Hemophilia, Genetic Disorder, India, Strategy

Corresponding Author
Mrs. Shabnam Mushtaq,
shabnammushtaq641@gmail.com

DOI
10.29121/shodhkosh.v5.i6.2024.4902

Funding: This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

1. INTRODUCTION

Given the ancient history of emigration and much diversified community layout in India exemplified by societal, geographical, lingual, and religious seclusion, are considered to be the primary causes of the country's hereditary variety (Basu et al.2016, 1594-1599; Mohanty et al. 2013, 33-42.). With the growing population, high fertility rate and biological isolation of many endogamous social groups and communities have also contributed to a relatively high prevalence of hereditary disorders in India. Every year approximately 495,000 infants are born with congenital deformities, 390,000 with (Glucose- Phosphate-dehydrogenase deficiency (G6PD) and 21,400 people with Down syndrome (Verma & Bijarni 2002, 192-196).

There are few comprehensive demographic statistics for other hereditary defects such heamophilia, cystic fibrosis, spino-muscular degeneration, myotonic degeneration, and fragile X dysfunction (Ananyeva et al. 2009,735-751). This

lack of data reinforces the idea that hereditary illnesses apart from haemo-globinopathies are sporadic, self-limiting problems for which preventative initiatives are not urgently needed.

However, in developing country like India, the burden of these disease is greater as compared to the western countries mainly due to the inadequate diagnostic, management and rehabilitation facilities (Balgir 2012, 3-13). As a result, many suspected cases of hereditary diseases remain undiagnosed or misdiagnosed in the region, causing patients to face a huge psycho-socio-economic crisis ailment (Basu et al .2016, 1594-1599). Despite the fact that hereditary disorders are infrequent and have a minimal prominence in India's national healthcare system, they cause enormous pain for both sufferers and their families (Wilkie et al. 2010, 391-400).

2. HEAMOPHILIA IN INDIA

According to some estimates, emerging nations including India are home to majority of the persons living with heamophilia. However, with many cases continue to remain unreported or under-diagnosed. As per the latest statistics of world federation of Haemophilia assessment, India is home to 13,448 individuals suffering from heamophilia. This figure is based on the information given by the Hemophilia Federation of India (HFI) (Balgir 2005, 1021-1026).

Hemophilia is an X-linked recessive genetic condition characterized by inadequate amounts of vital clotting proteins. The two highly prevalent types of heamophilia are *Haemophilia A* also known as factor VIII deficiency and *Haemophilia B* which is known as IX deficiency or as Christmas disease. which affects about 4 lac persons worldwide, is thought to impact 1 in 5000 males in its A and in its type B it affects about 1 in 30,000 live births (Chatterjee et al. 2010, 548-560). Both of its types have similar clinical symptoms and can show in modest, intermediate or extreme ways. If not managed immediately or prophylactically, the chronic type of haemophilia would be frequently marked by recurrent hemarthrosis, [1]which can progress to chronic crippling hemarthropathy[2] (Dye et al. 201, 81-90).

Directed at individuals with an afflicted birth or an ancestral background of the condition, hereditary intervention is the primary strategy for the treatment and management of hereditary ailments (Ananyeva et al. 2009, 735-751)

The World Federation of Haemophilia (WFH) regulations, which were released in 2005 and upgraded in 2012, offer doable advice for diagnosing and treating haemophilia. They primarily offer advice on how to handle challenges like musculo - skeletal problems, inhibitors, and outbreaks brought on by transfusions (Srivastava et al. 2020, 1-158)

Haemophilia treatment, unfortunately, continue to face numerous obstacles. Issues like accessibility of techniques and funds to ensure identification, and the range of treatment approaches for the condition and its consequences play a major role. Furthermore, financing may be discriminatory, unreliable, lacking, or otherwise impacted by governmental and administrative procedures (Ghosh and Shukla 2017, 451-452).

It is worthwhile to mention that the long-term medical disorders frequently face the continual difficulty of patient compliance towards medication regimes and follow-ups. According to estimates, up to 20 percent of haemophiliacs fail to take their recommended medications. This situation poses a challenge for a state to provide the best treatment because of different medical cost, recovery regimes, financial restrictions, and regional and societal variables particularly among the developing states. In case of developed state, having adequate supply of medication options, they are encumbered by increased expectations for consequences, along with a lack of supporting assistance to track adherence and results and establish a regular follow-up schedule (Jain et al. 2012, 911-913.).

The situation in India is quite concerning. Hemophiliacs unfortunately lack sufficient resources for management and detection of the disorder. The treatment of these patients are severely hampered by the absence of adequate testing and therapeutic resources as well as by the lack of knowledge about haemophilia among general practitioners and other experts (Lee et al. 2007, 391-394). For instance, it is difficult to find coagulation institutes in a consistent way. Ghosh et al. (2010) in his study reveal that lack of coagulation monitoring resources in the preponderance of district healthcare institutions and even medical institutions has an effect on how newer patients are diagnosed. It is very difficult to find resources for detecting and verifying inhibitors.

[1]*Hemarthrosis is a condition of articular bleeding that is into the joint cavity. This mainly occurs in a bleeding disorder such as hemophilia*

[2]*Hemarthropathy is a permanent and irreversible joint damage and is the most common complication of hemophilia that leads to disability.*

Resource oriented countries have developed a quick response mechanism to deal with hemophilic patients which include home therapy and early intervention of bleeding episodes. In these countries patients have easy access to specialized treatment centers as well as these centers provide training and awareness to hemophilic affected population. These two things are to be considered important component of comprehensive care. However in emerging nations budgetary constraints hinder the process of comprehensive care, in terms of not able to buy the sufficient amount of clotting factor. Moreover very few patients are able to purchase the treatment products even only for on-demand therapy. Besides these implications patients have very minimal access to treatment centers and comprehensive care (Pratap et al. 2020, 356-364).

3. THE WAY FORWARD

Therefore, it is challenging to compare the efficiency of treatment and develop support systems that take into account available capabilities (Gupta et al.2019, 190-199). Although there are various gaps in the treatment of haemophilia across different nations. It was suggested that the members of this community should assist each other in order to attain mutual interests (Pratap et al. 2020, 356-364).It is essential to abandon the search for a single ideal approach and, instead, benefit from the worldwide parallels and discrepancies in the provision of haemophilia management, developing the optimal strategy, adapted to every medical institution and patient group. There is a need for the establishment of a flexible and adaptive national standard of treatment with an emphasis on consistently high-quality care. The associations included nurses, physiotherapists, social workers, and patients (Marchesini, Morfini, and Valentino 2021, 221-235).

An integrated treatment approach is required that addresses all physical, psychological, and sociological needs of Patient with haemophilia and their caregivers. This approach is common in some developed nations like Canada.In addition, techniques like telemedicine should be employed to facilitate communication between patients and their caregivers. Patients and nurse coordinators should communicate over the phone frequently. Similarly outreach clinics in more patient-heavy smaller centers should be formed and. needy families should be identified and financially assisted by government-funded programmes.

Treatment based on empirical research should be incorporated in the field of haemophilia. Besides, that local context must be analyzed while providing the health care to these patients. Moreover, the care delivery system should be vigorous and accessible to all the sections of the society. It is also pertinent to mention that haemophilia health care delivery system must be globally collaborated with different organizations which will enhance the access to various treatment products around the globe (Stoffman et al. 2019, 39-48).

Home infusions are tracked through a national registry system in Canada. Patients electronically enter their information, which is then analyzed to determine whether the treatment plan is appropriate and to keep track of product supplies and usage. Maintaining registries is a must as it helps to understand the process of resource mobilization. These registries work as a road map for the government and different organization working for haemophilia to analyze the heamophilia scenario of the country and utilization of treatment products. Moreover, these registries are used to bring awareness among the people about the burden of the disease (Tahiraj 2014).

Lastly, a periodic independent assurance review is also necessary for the minimal accessible research institutes in order to boost their efficiency. The organizations working for the heamophilia must have to develop different programmes and policies which cater the need of these hemophilic patients. Besides these policies, it is advised to understand the actual disparities in heamophilia care across the various nations adopt those which are appropriate and can be considered as good practices. Though they possibly have not easy accessibility for component substitution, developing nations have created great physiotherapy and rehabilitation programmes. Even if the numerous investigations have sought to evaluate country-specific disparities in haemophilia management, relatively handful analyze haemophilia concern between healthcare systems that are financially imbalanced and the difficulties in providing adequate haemophilia care (Kar et al. 2014, 19-31).

CONFLICT OF INTERESTS

None.

ACKNOWLEDGMENTS

None.

REFERENCES

Ananyeva, Natalya M., Timothy K. Lee, Nisha Jain, Midori Shima, and Evgueni L. Saenko. "Inhibitors in hemophilia A: advances in elucidation of inhibitory mechanisms and in inhibitor management with bypassing agents." In *Seminars in thrombosis and hemostasis*, vol. 35, no. 08, pp. 735-751. © Thieme Medical Publishers, 2009.

Balgir, R. S. "Spectrum of hemoglobinopathies in the state of Orissa, India: a ten years cohort study." *The Journal of the Association of Physicians of India* 53 (2005): 1021-1026.

S Balgir, R. "Community expansion and gene geography of sickle cell trait and G6PD deficiency, and natural selection against malaria: experience from tribal land of India." *Cardiovascular & Hematological Agents in Medicinal Chemistry (Formerly Current Medicinal Chemistry-Cardiovascular & Hematological Agents)* 10, no. 1 (2012): 3-13.

Basu, Analabha, Neeta Sarkar-Roy, and Partha P. Majumder. "Genomic reconstruction of the history of extant populations of India reveals five distinct ancestral components and a complex structure." *Proceedings of the National Academy of Sciences* 113, no. 6 (2016): 1594-1599.

Chatterjee, Nirupama, Amit Mishra, Ravindra Soni, Hemant Kulkarni, Manju Mamtani, and Manisha Shrivasatava. "Bayesian estimates of the prevalence of β-thalassemia trait in voluntary blood donors of central India: a survey." *Hemoglobin* 34, no. 6 (2010): 548-560.

Dye, Danielle E., Kate J. Brameld, Susannah Maxwell, Jack Goldblatt, and Peter O'Leary. "The impact of single gene and chromosomal disorders on hospital admissions in an adult population." *Journal of community genetics* 2 (2011): 81-90.

Ghosh, Kanjaksha, and Rinku Shukla. "Future of haemophilia research in India." *Indian Journal of Hematology and Blood Transfusion* 33, no. 4 (2017): 451-452.

Ghosh, K., S. Shetty, and D. Sahu. "Haemophilia care in India: innovations and integrations by various chapters of Haemophilia Federation of India (HFI)." *Haemophilia* 16, no. 1 (2010): 61-65.

Gupta, Naresh, Abderrahmane Benbouzid, Meriem Belhani, Mohammed El Andaloussi, Khadija Maani, Yasser Wali, Soraya Benchikh El Fegoun, Hossam Ali Saad, and Johnny Mahlangu. "HAEMOcare: the first international epidemiological study measuring burden of hemophilia in developing countries." *TH Open* 3, no. 02 (2019): e190-e199.

Jain, Dipty, Khushnooma Italia, Vijaya Sarathi, Kanjaksha Ghoshand, and Roshan Colah. "Sickle cell anemia from central India: a retrospective analysis." *Indian Pediatrics* 49 (2012): 911-913.

Kar, Anita, Supriya Phadnis, Sumedha Dharmarajan, and Juhi Nakade. "Epidemiology & social costs of haemophilia in India." *Indian Journal of Medical Research* 140, no. 1 (2014): 19-31.

Lee, V. N., A. Srivastava, M. Nithyananth, P. Kumar, V. M. Cherian, A. Viswabandya, V. Mathews et al. "Fracture neck of femur in haemophilia A–experience from a cohort of 11 patients from a tertiary centre in India." *Haemophilia* 13, no. 4 (2007): 391-394.

Marchesini, Emanuela, Massimo Morfini, and Leonard Valentino. "Recent advances in the treatment of hemophilia: a review." *Biologics: Targets and Therapy* (2021): 221-235.

Mohanty, D., R. B. Colah, A. C. Gorakshakar, R. Z. Patel, D. C. Master, J. Mahanta, S. K. Sharma et al. "Prevalence of β-thalassemia and other haemoglobinopathies in six cities in India: a multicentre study." *Journal of community genetics* 4 (2013): 33-42.

Pratap, Rohan, Monali Misra, Varun N, Suman Morampudi, Anand Patil, and Jayachandra Reddy. "The existing scenario of haemophilia care in Canada and China-A review." *Hematology, Transfusion and cell therapy* 42, no. 4 (2020): 356-364.

Srivastava, Alok, Elena Santagostino, Alison Dougall, Steve Kitchen, Megan Sutherland, Steven W. Pipe, Manuel Carcao et al. "WFH guidelines for the management of hemophilia." *Haemophilia* 26 (2020): 1-158.

Stoffman, J., N. G. Andersson, B. Branchford, K. Batt, R. D'oiron, C. Escuriola Ettingshausen, D. P. Hart et al. "Common themes and challenges in hemophilia care: a multinational perspective." *Hematology* 24, no. 1 (2019): 39-48.

Tahiraj, Asllan. "Haemophilia care in Arabic countries compared to western European countries." PhD diss., lmu, 2012.

Verma, I. C., and S. Bijarnia. "The burden of genetic disorders in India and a framework for community control." *Community Genetics* 5, no. 3 (2002): 192-196.

Wilkie, Andrew OM, Jo C. Byren, Jane A. Hurst, Jayaratnam Jayamohan, David Johnson, Samantha JL Knight, Tracy Lester, Peter G. Richards, Stephen RF Twigg, and Steven A. Wall. "Prevalence and complications of single-gene and chromosomal disorders in craniosynostosis." *Pediatrics* 126, no. 2 (2010): e391-e400.