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Taking on a pandemic’s challenge: Emergent solutions from a single Hemophilia treatment center in North India

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ABSTRACT

Introduction: The global pandemic caused by SARS–COV-2 infection has raised several unique concerns in the bleeding disorders community. Although the risk of COVID-19 infection is not increased in patients with inherited bleeding disorders, the indirect effects of this infection are many.

Methods: A cross sectional survey was conducted among patients registered to our centre with inherited bleeding disorders. A web-based based questionnaire was developed and shared with patients and families.

Results: 120 patients/families answered the questionnaire completely. During the period of lockdown, many had bleeds that were left untreated due to either difficulty in travel or unavailability of treatment. The time to treatment ranged from 8 h to 15 days in those who had a bleed. 36 % faced financial difficulties and 40 % families reported losing their job or source of income during this period.

Discussion: Few solutions that emerged while treating patients during this period and recommendations are discussed. Even though haemophilia has been included under the essential health services and states mandated to continue treatment for these patients despite the global crisis, patients still face challenges in terms of transport and finance.

1. Introduction

The global pandemic caused by SARS–COV-2 infection has raised several unique concerns in the bleeding disorders community [1]. The direct effect of COVID-19 pandemic would be the risk of infection for people with hemophilia. Considering the contagiousness of this virus, living with the infection will soon be the norm than the exception [3]. In most immunocompetent haemophilic patients, the infection should have a similar course to that in non-haemophils. Sick COVID-19 haemophils risk the chance of bleeding during invasive procedures and mechanical ventilation. The indirect effects of COVID-19 are however more complex. There are concerns ranging from non-availability of clotting factors, poor access to care due to travel restrictions and fewer hospitals with facilities for hemophilia care such as factor support, physiotherapy or surgeries as many Hemophilia Treatment Centres (HTC) have been converted into COVID-only facilities [1]. There is also a fear psychosis related to COVID-19 which makes less people seek healthcare during this period. And finally, the economic burden of lockdown and loss of financial stability are what will affect patients most in resource-constrained settings. In the present study we examine the challenges faced by these patients and families with hemophilia and other inherited bleeding disorders with respect to COVID-19 pandemic.

2. Methods

A cross sectional survey was conducted among patients registered to our centre with inherited bleeding disorders. Our centre is a pediatric tertiary care teaching hospital located in North India in the National Capital Region. We cater to patients from Uttar Pradesh, Delhi and other adjoining states such as Uttrakhand, Bihar and Punjab. The questionnaire-based survey was administered over a period of 2 weeks from 15 to 30 June 2020 to patients attending the day-care clinic of the hospital. A simple Google-sheet based questionnaire was developed and shared with patients and families with hemophilia that was administered by the hemophilia counsellor directly to those patients who visited the hospital and through telephone to those who were at home after taking verbal consent. Patients were not called to hospital for the purpose of the study. In case the child was less than 12 years, one of the parents were interviewed. Institutional ethics approval was taken prior to the survey.

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3. Results

Out of 201 patients with inherited bleeding disorders registered at our centre, 135 patients/families answered the questionnaire. 15 questionnaires were incomplete and were not analysed; 120 were used for further analysis. Although ours is a pediatric institute, few adults receive treatment due to non-availability of treatment centres nearby. 41 were interviewed in the hospital where as rest were over phone.

3 girls were suffering from Von Willebrand Disease or rare bleeding disorders; rest were boys. Majority of patients were districts of western Uttar Pradesh for which our centre serves as a tertiary care facility. There were respondents from nearby states of Delhi, Haryana, Punjab, Bihar and Jammu and Kashmir. Median age of patients was 11 years (range 1–32 years). Diagnoses of patients who participated included severe haemophilia (95 %), moderate hemophilia (2%), Von Willebrand disease (2%) and Factor VII deficiency (1%). The median distance travelled by patient to reach this centre was 35 km and it ranged from 1 to 750 km.

3.1. COVID and hemophilia

Out of the 41 patients who visited the hospital for a bleed, only 2 has received any form of treatment (plasma/factor concentrate) prior for that bleed. The average duration of symptoms ranged for 8 h-15 days for this bleed. On being enquired about COVID-19, 119 out of 120 patients knew about the infection and replied that social distancing, mask, hand hygiene and staying at home can help them prevent the infection. On being enquired if COVID-19 infection had a direct bearing on hemophilia related bleeding, 49 out of 120 replied yes and they feared that contracting this infection would aggravate their bleeding phenotype.

During the lockdown period (Mid-March to Mid-June) in India, where schools and most offices had been shut, 83 out of 120 respondents had atleast one serious or life threatening bleed during this period. Serious bleed included hemarthrosis, muscle bleed, mucosal bleeding and life-threatening included intracranial, neck/throat bleeds and gastrointestinal bleeding [2]. 73 (60 %) felt that their bleeding phenotype was less frequent when compared to before whereas the rest had a similar pattern. 10 patients had an episode of life-threatening bleeding; 2 were intracranial, 2 were from gut (blood in stools) and 6 were genitourinary (blood in urine). Out of these 83 patients who had a major bleed, only 10 received any kind of treatment for it. The reasons for not being able to avail treatment were the fear of stepping out of house (11 %), travel restrictions (13 %), unavailability of public transport (15 %), restrictions by police to cross borders of states/districts (15 %), no factor availability (32 %) and that HTC was made into a COVID only hospital (35 %) respectively.

3.2. Treating hemophilia

Only 2/41 patients who experienced a bleed during this period could receive any form of treatment (plasma/factor concentrate) prior for visiting our centre for that bleed due to reasons mentioned before. 23 patients (19 %) had to visit more than 3 hospitals before treatment for hemophilia was possible due to either factor concentrate not being available or hospital being converted to COVID-only status. 14 patients had a physiotherapy session and 16 had an online consult with a physiotherapist during this period. 63 (52.5 %) however were continuing exercises which were taught earlier at home.

Out of these only 10 patients were on prophylaxis as our centre receives factor support from government for episodic treatment only. Those who were on prophylaxis were getting factor support through their employee insurance or were purchasing themselves. None of our patients were on World Federation of Hemophilia humanitarian aid program. 8 out of these 10 patients continued to have access to prophylaxis. 2 patients in our group were advised surgery during this period. One of them was performed whereas the other was postponed.

3.3. Economics of haemophilic families

33 families answered that there has been a change in their financial status since the onset of the lockdown. 44 patients/families feel that this financial difficulty has had a bearing on the treatment of the person with hemophilia. In 50 (41 % of respondents) families either one or more persons have lost their source of income during this period. 30 patients had to move to their native place due to difficulties encountered while living in towns. 20 families experienced financial difficulty to bring haemophilic patients to hospital for bleed.

3.4. Solutions and recommendations

Since travel was the major concern due to reduced access to travel and increased cost of travel the solutions that we considered subsequently are

a Counselling regarding COVID-19 and its effect on hemophilia patients. As immunity of haemophilic patients is intact, even when encountered with COVID-19 the response would be similar to other patients. There was no need for factor replacement for nasopharyngeal swab testing in case need arises. If admitted for this infection, a COVID hospital attached to their hemophilia treatment centre may be chosen so that prophylaxis/treatment support may continue uninterrupted during this period. Regular counselling has helped improve the fear of patients and visits to hospital for a bleed has improved subsequently.

b For patients who came with a bleed, an increase in dose of factor concentrate was considered. The usual treatment protocol of resource constrained settings with lower dose of factor support was switched to the dosing suggested for centres where resources were not a challenge. This helped us reduce further visits to the hospital except in major bleeds such as genitourinary and intracranial. For eg., the desired level of factor was raised to 40–60 u/dl instead of 10–20 u/dl which we regularly follow. For few patients, weekly visits were planned for factor support to enable physiotherapy without the fear of recurrence of bleed.

c For patients on prophylaxis (factor VIII/IX/emicizumab) treatment was continued with modification in schedule as permitted by the child’s activity since schooling and sports activities were suspended during this period.

d Home therapy would have been ideal in this situation; however, since we do not have access to prophylaxis for the majority of our patients, this was not possible in our scenario.

4. Discussion

The 2019 novel corona virus is currently causing the severe outbreak named COVID-19 in almost all countries of the world and thus has been labelled a global pandemic [3]. Although the risk of infection in an immunocompetent host such as a patient with haemophilia with no co-morbidities is not higher than normal population, the chance of bleeding may be higher if patient is admitted with severe COVID infection. The indirect effects of the infection are however very far reaching and with significant consequences. In other patient There are concerns ranging from non-availability of clotting factor, poor access to care due to travel restrictions and fewer hospitals with investigatory and treatment facilities including factor support, physiotherapy and surgery as many have been converted to COVID only hospitals to fear of stepping out of home even in case of bleed. Another major challenge is for patients supported through the World Federation of Hemophilia humanitarian aid program which will now reduce due to restrictions on air travel [4].

There is a fear psychosis related to COVID which makes people seek care less during this period. This was observed in our series of patients also. The economic burden of lockdown and loss of job opportunities are...
probably what will affect patients most in resource constrained settings. Having to deal with a chronic high maintenance disease in the midst of an economic crunch can be disheartening and extremely challenging to patients and families alike. The efforts from central and state government have been very supportive in this regard.

Hemophilia has been included under the essential health services and all states have been mandated to continue treatment for these patients despite the global crisis. Despite the fact that 70% of patients interviewed for this survey had a major bleeding during lockdown, only about half of them could reach a centre for treatment. The difficulty to travel in order to reach the treatment centre and fear of COVID were the main reasons for not having received treatment. Many also had severe financial constraints that made it difficult to afford private vehicles to reach the hospital.

Eventually we need to identify solutions and move on with this infection amongst us [5,6]. Among the solutions, counselling regarding COVID appropriate behaviour, avoidance of panic, need for adherence to physiotherapy continue to help us navigate this period. Although prophylaxis was not an option for many, a higher dose of factor support for on demand period and regular visits for factors helped prevent a re-bleed. Having to deal with a chronic high maintenance disease in the midst of an economic crunch can be disheartening and extremely challenging to patients and families alike.

Author contribution

NR prepared the manuscript. NR, RS and MR managed patients. SS was involved in the diagnosis. All authors were involved in writing the manuscript.

Declaration of Competing Interest

The authors report no declarations of interest.

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