



Commentary

Challenges in successful management of haemophilic knee arthropathy: Our experience in a tertiary centre

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ABSTRACT

Background: Haemophilia is characterized by a partial or complete deficiency of clotting factor VIII or IX. The repeated bleeding episodes into the joints contributes to subsequent chronic arthropathy which debilitates the patient and affects the quality of life. There are lot of hurdles in successful management of such patients. We present our experience in dealing such patients.

Materials and Methods: 24 patients (27 knees) with haemophilic knee arthropathy were included. The pre-operative VAS (Visual Analogue Scale) and KSS (Knee Society Score) were assessed. All underwent total knee arthroplasty with stemmed constrained prosthesis. Postoperatively VAS and KSS were analysed at the end of 12 months.

Results: The mean preoperative VAS was 8.2 (Range: 7–10), mean KSS clinical score was 39.9 (Range: 12–61) and mean KSS functional score was 51.4 (Range: 20–70). The mean postoperative VAS was 2.7 (Range: 2–4), mean KSS clinical score was 70.5 (Range: 61–80) and mean KSS functional score was 74.2 (Range: 60–80). There was statistical significant difference between preoperative and post-operative scores. At the end of 12 months, 17 patients (63%) had good outcome, seven patients (26%) had fair and three patients (11%) had excellent outcome as per the KSS scores.

Conclusions: Total knee arthroplasty has good outcome with respect to clinical as well as functional outcome in haemophilic arthropathy. Although the results and outcome of total knee arthroplasty in haemophilic knee arthropathy may still be inferior to the results seen in a normal individual, a multidisciplinary approach yields a better functional and quality of living in such haemophilic patients.

1. Introduction

Haemophilia is a coagulation disorder which follows X-linked recessive pattern of inheritance. It is characterized by the deficiency of clotting factors VIII (FVIII, haemophilia A) or factor IX (FIX, haemophilia B). The most common form is factor VIII deficiency, or haemophilia A, which comprises approximately 80% of cases.^{1,2} The deficiency of clotting factors results in spontaneous and repeated bleeding into the muscles and joints, which is the most common clinical manifestation of haemophilia. The most commonly affected joints in haemophilia are knee, ankle, elbow and hip joint.¹ The articular problems in haemophilia affected patients begin from their early age itself, recurrent injuries leading to recurrent hemarthroses, repetitive synovitis leading to flexion deformities, hypertrophy of the growth plates, alteration of the bony architecture and cartilage finally leading to end stage haemophilic arthropathy.^{3,4} Due to chronic inflammatory reaction, at microscopic

level there is ongoing deposition of hemosiderin, chemical inflammatory mediators like cytokines that initiates neoangiogenesis which in turn results in cartilage damage by causing chondrocyte apoptosis.³

Arthroplasty is the treatment of choice in end stage haemophilic arthropathy.^{4–6} The purpose of this study is to put forward our experience and challenges in dealing with such patients with end stage knee arthropathy.

2. Materials and methods

This study was conducted in our hospital which caters to a large number of haemophilic patients. Our case series is based on a 5 year chart review between year 2012–2017 of orthopaedic and joint replacement registry data with diagnosed cases of haemophilia with orthopaedic concern. Both the types of haemophilia were included in our

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Table 1

Table showing Pre-operative Range of movement (ROM), Knee society score (KSS) clinical, functional score and Visual analogue Scale (VAS).

Serial number	AGE	ROM	KSS CLINICAL	KSS FUNCTIONAL	VAS
1	27	41–45	28	55	8
2	27	46–50	28	55	7
3	29	61–65	53	60	7
4	32	41–45	21	45	9
5	34	36–40	18	45	9
6	26	51–55	49	60	7
7	29	61–65	41	50	7
8	36	46–50	35	45	9
9	30	61–65	53	55	8
10	35	51–55	60	60	9
11	35	56–60	60	60	8
12	37	46–50	30	45	9
13	31	61–65	51	70	8
14	27	71–75	53	65	8
15	24	56–60	60	70	7
16	39	36–40	23	20	9
17	38	51–55	30	45	9
18	38	46–50	30	45	8
19	41	31–35	12	30	10
20	33	61–65	61	60	8
21	30	61–65	53	55	8
22	32	41–45	21	45	9
23	39	36–40	23	20	9
24	26	51–55	49	60	7
25	33	56–60	60	60	8
26	32	41–45	21	45	9
27	33	61–65	61	60	8

Table 2

Table showing Post-operative Range of movement (ROM), Knee society score (KSS) clinical, functional score and Visual analogue Scale (VAS).

Serial number	AGE	ROM	KSS CLINICAL	KSS FUNCTIONAL	VAS	outcome
1	27	61–65	71	80	3	good
2	27	71–75	71	80	3	good
3	29	76–80	76	80	2	good
4	32	66–70	64	60	4	fair
5	34	56–60	65	70	3	fair
6	26	66–70	72	80	3	good
7	29	71–75	73	70	3	good
8	36	61–65	61	80	3	fair
9	30	76–80	64	70	3	fair
10	35	66–70	72	80	2	good
11	35	66–70	72	80	2	good
12	37	66–70	63	70	3	fair
13	31	81–85	75	80	2	good
14	27	81–85	80	80	2	excellent
15	24	76–80	74	80	2	good
16	39	51–55	72	70	3	good
17	38	66–70	74	80	3	good
18	38	61–65	74	80	3	good
19	41	61–65	66	70	3	fair
20	33	76–80	80	80	2	excellent
21	30	76–80	72	70	3	good
22	32	66–70	72	60	3	good
23	39	51–55	71	70	3	good
24	26	66–70	72	80	3	good
25	33	66–70	72	80	2	good
26	32	66–70	64	60	4	fair
27	33	76–80	80	80	2	excellent

study who were symptomatic and had grade 3, grade 4 knee arthropathy as per Modified Arnold Hilgartner grading. Patients who had Von willebrand's disease and other bleeding disorders were excluded. There were 24 haemophilic patients with symptomatic knee arthropathy knee included in our study. 18 patients were suffering from haemophilia A and six patients were suffering from haemophilia B. All patients were managed as per standard WHO (World Health Organisation) protocol



Fig. 1. Lateral radiograph of the knee joint with prosthesis insitu with patella fracture (yellow colour pointer).

for haemophilia by the haematology department. The preoperative VAS (Visual Analogue Scale) and KSS (Knee Society Score) were assessed and compared with VAS and KSS at the end of 1 year post-operative period.

Ethical committee clearance and standard procedure consent were obtained from the patients.

2.1. Operative workup and protocol

After obtaining the pre anaesthetic check-up, all patients were admitted under the haematology department. Patients factor levels and presence of any factor eight inhibitors in the blood were assessed prior to surgery. Accordingly patient were started on factor 8 or factor 9 injections to build up the levels of factors beyond 80%. On the day of surgery, the case is accepted for surgery only if the circulating factor 8 or factor 9 levels in the body are beyond 80%, if not a repeat injection of factors is given and bloods are repeated again for factor levels. Once the factor levels are above 80% case is taken up for surgery.

All cases are done under general anaesthesia with adequate back up of PRBC (packed red blood cells), FFP (fresh frozen plasma), platelets and factor 8/factor 9 injection. In case of patients with factor 8 or 9 inhibitors positive status, such patients needed Factor inhibitors bypassing agents. All cases were done under the effect of tourniquet and also tranexamic acid.

Standard anterior midline, medial Para patellar approach to knee joint was used in all cases.

The tibial and femoral cuts were made as per the intramedullary jigs. All patients underwent total knee arthroplasty with constrained stemmed prosthesis. Tourniquet was deflated after the polymerisation of cement and all the bleeders were cauterised before closure of wound. No drains were used in any of our cases. Wounds closed in layers with



Fig. 2. A: Clinical photographs showing the knee flexion deformity on table.
 B: Clinical photograph showing the equinus deformity of the ankle.
 C: Clinical photograph of the patient showing the deformity of the knee and ankle.

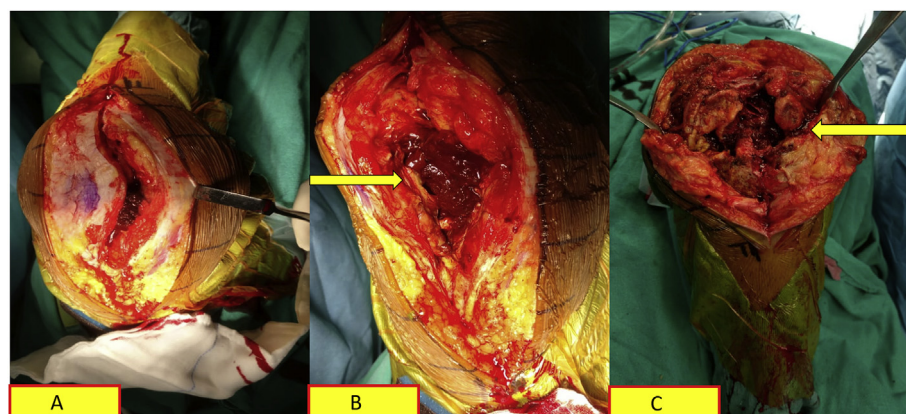


Fig. 3. A and B: Clinical photographs showing extensive haemosiderin laden soft tissue within the knee joint (yellow pointer).
 C: Clinical photograph showing the extensive soft tissue contractures (yellow pointer).

vicryl sutures and skin closed with staples. Non adhesive dressing was used in all cases with above knee compression bandage.

Postoperatively the factors levels were repeated on daily basis and factor injection given accordingly. Post-operative day 2 onwards patients were put on CPM (continue passive mobilisation) twice daily.

Walker assisted weight bearing is initiated once the patients were pain free. All patients were kept inpatient till suture removal.

3. Results

We included 24 patients in our study (18 patients: Haemophilia A, 6 patients: Haemophilia B). Of the 24 patients, three patients underwent bilateral total knee arthroplasty in the same sitting and the rest 21 patients underwent single side knee arthroplasty. Only Five patients underwent patella resurfacing as the rest 19 patients had thinned out patella. Three patients were HBsAg positive, one patient was HCV positive and one patient was positive for HIV. Three patients had liver cirrhosis secondary to viral hepatitis. One patient was factor 8 inhibitor positive for whom preoperatively and peri-operatively factor 8 inhibitor

bypassing agents (FEIBA) had to be given. One of the patients had repeated low haemoglobin post operatively and also had episode of malena due to bleeding from esophageal varices which was managed with endoscopic oesophageal variceal banding. One of them sustained a patella fracture three months post operatively which was managed conservatively. None of the patients had any postoperative infection however three patients had recurrent hemarthrosis perioperative period which was managed with factor supplementation, ice pack and delaying the mobilisation protocol.

All patients were followed up at 12 months postoperatively.

The mean preoperative VAS was 8.2 (Range: 7–10), mean KSS clinical score was 39.9 (Range: 12–61) and mean KSS functional score was 51.4(Range: 20–70). The mean postoperative VAS was 2.7(Range: 2–4), mean KSS clinical score was 70.5 (Range: 61–80) and mean KSS functional score was 74.2(Range: 60–80). Three patients had excellent result as per KSS score (80–100), 14 patients had good results (70–79) and seven patients had fair results (60–69). None of the patients had poor results (< 60) (Table 1). There was statistically significant difference in the pre and post-operative KSS score. 17 patients (63%) had

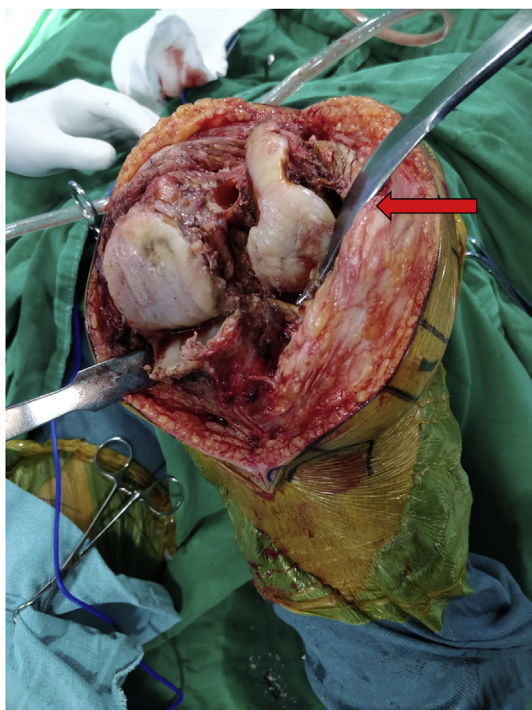


Fig. 4. Clinical photograph showing the thinned out patella being retracted with a Hohmann retractor (red pointer).

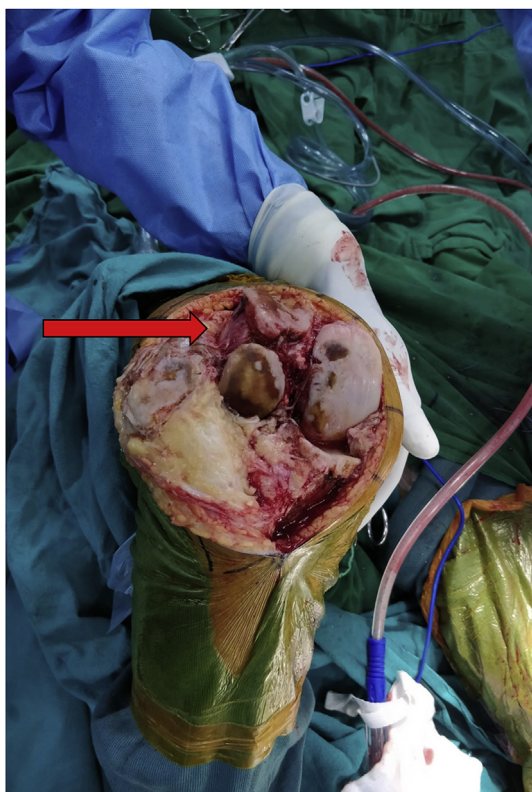


Fig. 5. Clinical photograph showing the altered bony anatomy. Red pointer pointing the lateral femoral condyle which is hypoplastic and worn out completely.

good outcome, seven patients (26%) had fair outcome and three patients (11%) had excellent outcome as per the KSS scores (Tables 1 and 2).

4. Discussion

Total knee replacement has been the gold standard treatment for end stage haemophilic arthropathy providing pain free, better functionality and quality of life.⁷ However due to end stage arthropathy, extensive soft tissue fibrosis, altered bony architecture and severe deformities, total knee replacement becomes even more challenging than the conventional total knee arthroplasty.⁸ All of the above mentioned reasons contribute to decreased preoperative range of movements which in turn affects the postoperative functional outcome.^{9–11} To begin with a preoperative range of movement of 50° or less is considered to have significant impact on the functional status of the patient and a range of movement of 65–90° is needed to carry out the activities of daily living.^{8–11} In our series of 27 haemophilic patients, 12 patients (45%) of the patients had a preoperative ROM of less than 50° and postoperatively 20 patients (75%) had ROM ranging 65–85°. We also documented a significant decrease in postoperative VAS score and improved postoperative KSS score. Despite the restricted ROM postoperatively, 17 patients (63%) had good outcome, seven patients (26%) had fair outcome and three patients (11%) had excellent outcome as per the KSS scores which was comparable to other studies.^{8,10,12}

Based on our experience and findings, postoperative outcome of arthroplasty in haemophilia may not be as good as the outcomes seen in regular cases but nonetheless the reduction in pain and improved functional outcome really makes a big impact on the activities of daily living in a debilitated haemophilic arthropathy patient.^{5,7,13}

None of our patients had any surgical site skin breakdown, surgical site infection, periprosthetic joint infections or a periprosthetic fracture postoperatively. One of our patients sustained a patella fracture postoperatively following a trivial trauma which went on to heal uneventfully with conservative management (Fig. 1). Finally here we sum up the various challenges faced by us in dealing with haemophilic patients under these following headings.

4.1. Medical challenges

Repeated blood transfusions increases the chance of development of inhibitors to factor 8. Treating such factor 8 inhibitor positive patients is extremely costly and FIEBA is not freely available making it even more challenging in treating such cases.

Repeated blood transfusions also make the haemophilic patients susceptible to HIV, HBsAg, and HCV etc which not only causes chronic hepatitis but also adds an additional risk to the operating surgeon making it even more difficult to operate on such cases.

Regular monitoring of the factor levels pre and perioperatively and also maintenance of at least 50% of factor levels postoperatively is mandatory to avoid bleeding tendency and hemarthrosis.¹⁴ This mandates an availability of a well organised lab facility and blood bank facility with adequate back up of factors, blood and blood products to avoid any catastrophic consequences perioperatively.

4.2. Surgical challenges

All cases are done under general anaesthesia. Complex deformities in hip, knee and also sometimes in ankle joint results in improper positioning of the patients (Fig. 2). Fixed flexion deformity, soft tissues contracture and haemosiderin laden soft tissues makes it even more challenging intraoperatively (Fig. 3). Resurfacing in thinned out patella may increase the chance of patella fracture and also one has to be very careful on table while everting the patella for better exposure (Fig. 4). Hence it is advisable to retract the patella rather than everting it. Altered articular surface and poor bone stock makes it difficult to place the jigs and prepare the bone cuts (Fig. 5). Extramedullary jigs may result in erroneous cuts, we advise intramedullary jigs for better accuracy of the tibial and femoral bone cuts. Tibia preparation to be done first in order to have better access to distal femur (Fig. 6). The use of

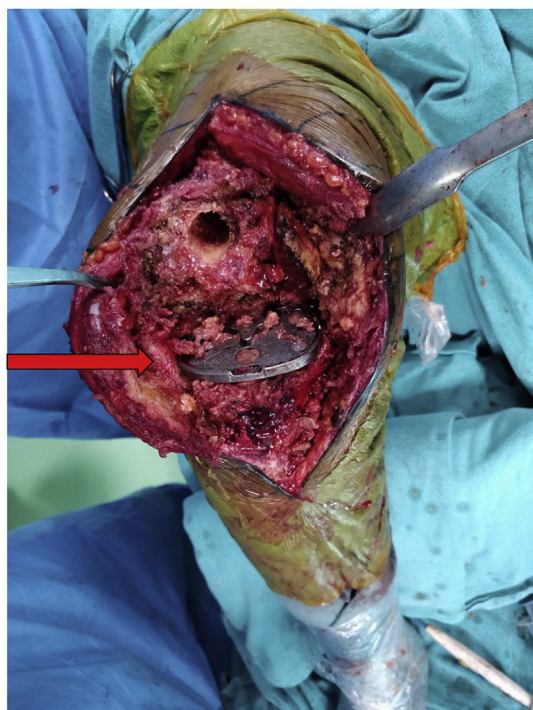


Fig. 6. Clinical photographs showing the preparation of the tibial surface before the femoral surface (red pointer).



Fig. 7. Post-operative radiographs with constrained stemmed prosthesis.

constrained stemmed prosthesis (Fig. 7).

Postoperatively serial monitoring of the factor assays and haemoglobin. Surgical site oozing and repetitive hemarthrosis will delay in the rehabilitation. Aggressive rehabilitation is detrimental resulting in hemarthrosis hence a slow and gradual rehabilitation protocol has to be followed. The use of continuous passive mobilisation (CPM) in the initial few days yields a better and gradual rehabilitation in these patients. Lastly possibilities of delayed wound healing and late surgical site infections can cause poor functional outcomes. The substantial post-operative improvement in quality of life has to be weighed against all the risks of operation. Successful management of haemophilic arthropathy needs a multidisciplinary approach. Hence we would like conclude that the post-operative outcome in haemophilic knee arthropathy may not be as good as the outcomes seen in regular cases but there is significant improvement in functional outcome and quality of life.

Declaration of competing interest

There are no conflict of interest.

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