More coordinated education and management of haemophilia patients required in India

A significant proportion of the Indian population is affected by hereditary defects in one or more of the clotting factors. Although effective therapy has been made available for haemophilia in the last 40 years, many issues remain unresolved regarding the management of this condition, particularly with reference to doses and duration of factor replacement therapy for different types of bleeding, immune tolerance induction, and surgical prophylaxis. In the developed countries like USA and Sweden, it is now possible for a child with haemophilia receiving adequate treatment to live a near normal life. An accurate diagnosis is quickly established, the family is educated on disease management and the child is put either on prophylactic factor replacement or on-demand replacement remedies taken at home. Prior to the 1960s, when no comprehensive care was available, individuals with haemophilia suffered a similar fate worldwide. Severe joint disabilities appeared in early teens and most patients died before their maturity. Haemophilia was treated primarily with fresh blood transfusions and as a result haemophilia associations were established for the purpose of recruiting the donors. The advent of cryoprecipitate in 1964 and subsequent development of clotting factor concentrates dramatically increased clinical management options. As concentrates could be easily stored, administered at home and carried with patients during travel, patients began to adopt a practice of home therapy. In USA, early treatment of bleeding episodes and home therapy quickly evolved as the primary management option. Training and education of patients about disease management became necessary with the increasing popularity of home therapy. Specialized centres soon delivered services to meet these needs. During the past 20 years, the cost of treatment rose exponentially (5–10 times). Today, the cost of optimum care for haemophilia is beyond the reach of haemophilia community so that others, whether from government or private insurance, must bear the expense. As a result, the individuals with haemophilia must continually convince others to accept this financing responsibility. Health is a human right, which has also been accepted in the Constitution. Its accessibility and affordability has to be insured. According to the recent reports from various agencies, the health sector in India has become a Rs 25,000-crore industry. With the entry of various private insurance companies now the customers have choice of buying this insurance from 16 insurance companies. However, to make haemophila treatment affordable in India we have to work according to the World Federation of Hemophilia guidelines in association with Haemophilia Federation of India (HFI). Some collective effort from the government, private health care industry and insurance companies should collectively support haemophilia community. The major problems with regard to haemophilia care in India are inadequate knowledge, lack of facilities for a proper laboratory diagnosis, and inadequate supply of an affordable and safe clotting factor. Due to lack of awareness, education and counselling, many patients do not come forward to give history of bleeding. Other factors like social stigma and lack of parent support are equally responsible. We report here a case series of five children presenting with massive bleed in post-operative/post-interventional period which were referred to us from 2008 to 2009. To our knowledge such case series are not reported in Indian paediatric/medical literature. Thus, there is a need for awareness regarding this issue among treating doctors.

Incidence of bleeding disorder amounts to about one in 10,000 births in total population. With an occurrence of von Willebrand's disease (VWD) as 1–2% of the total population and the haemohilia as 1 per 5000 male births, VWD and haemophilia are the most common bleeding disorders[1–6]. Consanguinity being very high in north Karnataka, the bleeding disorder may be more than projected[7]. An appropriate history is helpful in evaluating the type of bleeding disorder. Age of onset, frequency and severity of each bleeding complaint should be determined, and an extensive family history and medication history should be obtained before any intervention. Laboratory studies that assist in confirming the type of bleeding disorder include complete blood count, platelet count and

Table 1. Case profile of different bleeding disorder cases

Case no.	Age (years)	Sex	History given by doctor/patient	Education status of parents	Type of bleeding disorder	Intervention/ surgery	Post-op complication
1	18	M	None	Educated	Haemophilia A	Tooth extraction	Airway compression secondary to neck haematoma
2	3	F	None	Uneducated	VWD (von Willebrand's disease)	SICS	Conjuctival and sub conjuctival haemorrhage
3	7	M	None	Uneducated	Haemophilia A	Splint and tractions	Haemarthrosis
4	13	F	None	Uneducated	VWD	Joint aspiration	Septic arthritis
5	4½	M	None	Uneducated	Haemophilia A	Tooth extraction	Haemorrhage from soft palate

platelet functions prothrombin time, activated prothrombin time and factor estimation[8]. In the present study, mean age of the patients was 9.1 ± 5.6 years. Sixty per cent of the cases were males and the rest were females. Only one patient was educated whereas 80% parents were uneducated and were not fully aware about the disease. Eighty per cent of the cases were known cases of bleeding disorders (3 haemophilia and 1 VWD) whereas one was diagnosed after admission. None of the patients had provided a proper past history of bleeding episodes to the doctor nor any attempt was made by doctor to elicit this relevant history. In our study, 100% patients had complication following surgical intervention and in one case it was life threatening. None of the parents were educated regarding the disease and the first aid nor did they attend any awareness camp. In our case series of five patients the doctor did not suspect or elicit proper history of bleeding disorder in the patients. None of the patients underwent investigations for bleeding profile prior to surgery. HFI is active since 1983 and sponsoring treatment and education of children with haemophilia. Efforts of HFI, awareness and following the WFH guidelines in India are small steps towards harmonizing care of the people with bleeding disorder in the country until evidence-based practice is possible.

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On phylogenetic classification of fungi *sensu lato*: are we closing doors for morphotaxonomists?

Phylogeny remained speculative for very long, never had any role in classification and often formed the last chapter of mycological textbooks[1,2]. With the emergence of molecular techniques, suddenly it is taking the front seat. The other criteria employed for elucidation of fungal classification and rearrangement of various taxa/groups other than morphology and ontogeny include biosynthetic pathways, genetic basis, scanning electron microscope (SEM) for surface ornamentation, transmission electron microscope (TEM) for internal structure and presence/absence of organelles, cell wall composition, etc. Apart from beautiful diverse morphological characters for which fungi are known, taxonomists also used host association (parasitism, saprophytism/commensalism), serological reactions, anamorph–teleomorph connections and sexuality as parameters in taxa differentiation.

Fungi play an important role in ecosystem functions[3], therefore we need documentation/classification of these. Thus we enter the field of taxonomy, where the taxonomists play a central role with a herculean task at hand while 'inventorying the biodiversity of this planet'. Hawksworth estimated 1.5 million species of fungi based on 1 : 6 ratio of plant and fungi of the most extensively studied piece of land, the British Islands. But later he revisited his conservative estimate with a more sound basis of known plant species (270,000), the vast number of unknown insect species (5–30 million), the increasing number of fungi plant ratio in specific geographical regions and the possibility of more fungi in tropical/polar regions than in temperate regions and arrived at the estimate of 9.9 million species[4]. The large number of devoted morphotaxonomists could discover only 100,000 species of fungi. Tropics contain more than 50% of biodiversity, including fungi, but have less number of mycologists[5–6].

A glance through the recent mycological journals *Mycological Research* and *Mycologia* reveals that the entire traditional fungal classification is like an Iranian carpet turned upside down due to the phylogenetic/molecular analysis based classification. A few examples of groups will clarify this statement. In fact, almost all the groups have been phylogenetically reshuffled/rearranged. The traditional aplanospore/zygospore (presence or absence) forming fungi, earlier placed in Zygomycota now treated in subphylum Mucoromycotina, Kickxellomycotina, Zoopagomycotina and Entomophthoromycotina of fungi as *insertae sedis*[7] (not placed in any phylum). In the phylum Ascomycota[7,8], the order Erysiphales earlier treated in Pyrenomycetes now treated in Leotiomycetes and Meliolales earlier in the same class as Erysiphales are now placed in Sordariomycetes *inc. sed.* Braun and Takamatsu[9] as a result of rDNA ITS sequences synonymized well-known teleomorph morphology based distinguishable genera *Uncinula* (appendages curved apically), *Microsphaera* (appendage branched apically) and *Sphaerotheca* (appendages myceloid