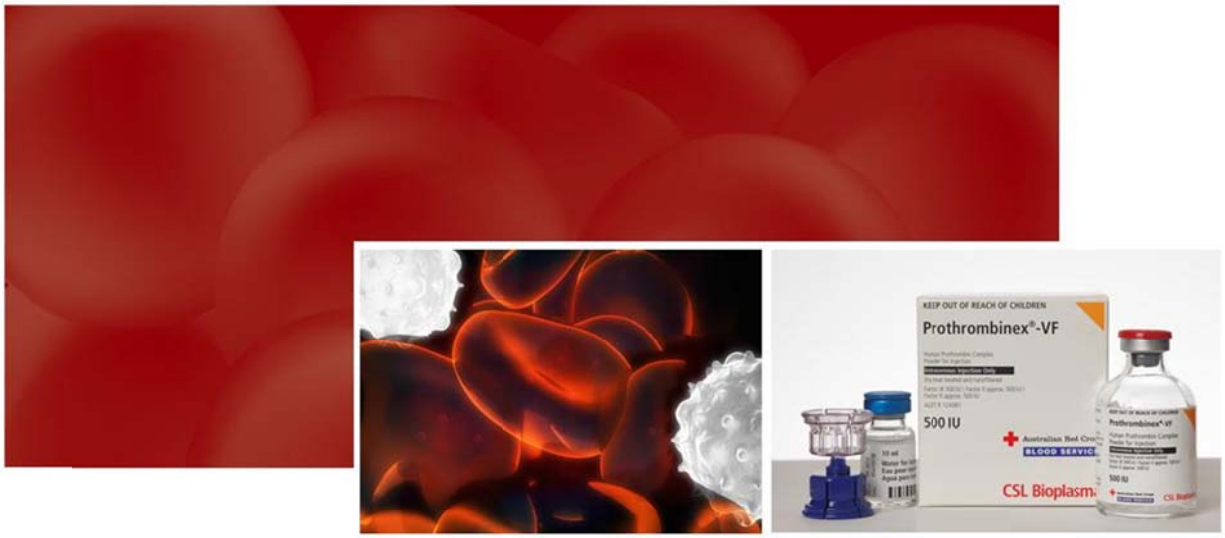

ABDR

Australian Bleeding Disorders Registry

Annual Report

2009-2010

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1.1. Glossary (main terms)

Abbreviation	Meaning
ABDR	Australian Bleeding Disorders' Registry
AHCDO	Australian Haemophilia Centres Directors' Organisation
DDAVP	Desmopressin (1-deamino-8-D-arginine vasopressin, abbreviated DDAVP) a derivative of the antidiuretic hormone, used to treat patients with Haemophilia A and von Willebrand disease. It does not come under the national blood agreement funding arrangements and its use in not recorded in the NBA's issues database.
HFA	Haemophilia Foundation Australia
HmA	Haemophilia A (Factor VIII deficiency)
HmB	Haemophilia B (Factor IX deficiency)
HTC	Haemophilia Treatment Centre
IDMS	Integrated data management system – The NBA's integrated data management system.
NBA	National Blood Authority
PWBD	People with a bleeding disorder
vWD	von Willebrand disease

2.

Executive Summary

2.1. Purpose

This document draws data from the Australian Bleeding Disorders Registry (ABDR) and other National Blood Authority supply and contract sources to provide an integrated view of current clinical and demographic information on people with bleeding disorders in Australia and the resultant demand for clotting factor products.

It has been developed through the close collaboration of all stakeholders involved in the management and governance of the ABDR, namely:

- Australian Haemophilia Centre Directors' Organisation (AHCDO)
- Haemophilia Foundation of Australia (HFA) and
- National Blood Authority(NBA).

2.2. Key observations

The data contained in the Report shows:

- the number of patients registered increased by 289 from 2008-09 to 2009-10;
- the number of patients with severe diagnosis increased by 28;
- there was no significant change in the proportion of people in the register treated between current data and earlier versions of the ABDR;
- there was a significant increase in the number of people in the register with Factor XI deficiency, albeit the totals are small, and those with von Willebrand disease (vWD); and
- there was an increase in the proportion of people in the register who were females.

These results are tempered by some data quality issues. Some records are incomplete and some product use is not recorded. In some areas inconsistent definitions appear to be used for some fields.

This report represents the first analysis of the ABDR data since a redevelopment in 2008 and allows comparison of data input by jurisdiction to improve collection in deficient areas. The collation of these data also allows some key insight into the way in which product is managed, for example, there is

- variability in dosage regimes by jurisdiction; and
- variability in completeness of data input by jurisdiction and by Haemophilia Treatment Centre (HTC)

3.

Background

3.1. What are bleeding disorders?

3.1.1. Haemophilia

Haemophilia occurs in 1 in 6,000-10,000 males internationally.

In Australia there are approximately 2,300 people with varied degrees of severity of this condition. There are 2 types of Haemophilia:

- Haemophilia A (classical Haemophilia) is the most common type and caused by a deficiency of blood clotting factor VIII.
- Haemophilia B (Christmas Disease) is due to a deficiency of blood clotting factor IX.

The deficiency in clotting factor produces a wide range of bleeding episodes, usually into the joints, muscles or internally possibly affecting vital organs. These bleeding episodes, or "bleeds", may occur spontaneously, or as a result of trauma or injury. The bleeding is stopped by infusion of the appropriate clotting factor by intravenous injection. Over a period of time bleeding into joints and muscles can cause permanent damage such as arthritis, chronic pain and joint damage requiring surgery.

With appropriate treatment haemophilia can be managed effectively.

Haemophilia is an inherited condition and occurs in families, however in one-third of cases it appears in families with no previous history of the disorder. Haemophilia is due to a mutation in the Factor VIII or factor IX gene which is on the X chromosome. The haemophilia gene is passed down from parent to child through generations. Some women and girls who carry the haemophilia gene may also experience bleeding problems.

3.1.2. von Willebrand disease

von Willebrand disease (vWD) is a related bleeding disorder which affects both men and women. This disorder is more common and is caused by a deficiency and/or malfunction of von Willebrand factor.

Table 1 Major bleeding disorders and their cause

Disorder group	**Cause**
Haemophilia A	Deficiency in factor VIII
Haemophilia B	Deficiency in Factor IX
von Willebrand Disease	Deficiency in von Willebrand factor
Other factor deficiencies	Deficiency in other coagulation factors.
Platelet Disorder	Deficiency in effective platelets

3.1.3. Severity

Haemophilia A and B are classified according to their severity as this informs the treatment regimens required. The definitions of severity that are applied within the ABDR are listed overleaf. Definition of severity of vWD and other coagulation factor deficiencies is variable.

Table 2 Severity and the concentration of clotting factors[1]

Severity	Concentration of Clotting Factor	Typical Bleeding Picture
Severe	<0.01 IU/ml (<1% of normal†)	Frequent bleeding episodes common, predominantly into joints & muscles. Bleeding can occur spontaneously or after minor injury.
Moderate	0.01 – 0.05 IU/ml (1–5% of normal)	Can bleed after minor injury. May have joint bleeding. Severe bleeding with trauma, surgery, invasive procedures.
Mild	>0.05 – 0.40 IU/ml (5-40% of normal)‡	Spontaneous bleeding does not occur. Bleeding with major trauma, surgery, invasive procedures.

Notes † Normal concentration of factor VIII or IX is defined as 100% or one unit of factor VIII activity per ml of plasma - 100 U/dL (Kasper 2004).
‡ levels of FVIII above 40% are considered sufficient for normal haemostasis.

3.1.4. Treatment of bleeding disorders

Mild conditions may require no treatment or treatment only under special circumstances such as surgery. More severe conditions may require regular interventions. Treatment may occur in a hospital or other medical facility, or at home. The treatments may be regular and preventative (prophylaxis), or on demand (when a bleed occurs).

Often the treatments involve providing replacement for the missing or defective clotting factors. Products used include plasma derived and recombinant clotting factors, cryoprecipitate and Desmopressin (1-deamino-8-D-arginine vasopressin, abbreviated as DDAVP). In some patients, their therapy is complicated when their bodies develop inhibitors that destroy the replacement clotting factors and other treatment is necessary.

3.2. Treatment arrangements in Australia

The majority of people with these conditions are treated at Haemophilia Treatment Centres (HTCs) which are specialist centres that provide comprehensive care to people with haemophilia and other coagulation deficiencies. The comprehensive care model ensures that preventative and general treatment on the complex aspects of haemophilia are given in a co-ordinated way by a multi-disciplinary team within the one centre.

Haemophilia Centres were established under a decision by the Australian Health Ministers Advisory Council in 1998 to provide a leadership role within their hospital, city and outlying areas to ensure optimal care and an equitable distribution of professional and therapeutic resources, together with responsible record-keeping. Specific roles of these Centres proposed by this decision are defined in Appendix A.

[1] Modified from Bolton-Maggs, PH & Pasi, KJ 2003, 'Haemophilias A and B', Lancet, 361 (9371), pp. 1801–1809. See also: White GC et al. Definitions in Hemophilia: Recommendation of the Scientific Subcommittee on Factor VIII and Factor IX of the Scientific and Standardization Committee of the International Society on Thrombosis and Haemostasis. *Thromb Haemost* 2001;85:560.

The manner in which these functions are delivered varies between states including the degree to which they:

- operate within a centralised compared to distributed treatment models;
- focus on treating paediatric or adult patients compared to treating all age groups; and
- have relatively small numbers of patients compared to relatively large numbers – commonly based on geography.

There are also some patients that receive product from clinicians not associated with an HTC. The proportion of product that is used in these circumstances varies across jurisdictions and there is therefore some variability in the data capture rate within jurisdictions. Accordingly, data on total volume of products recorded from the ABDR may not be consistent with data provided by the NBA from other sources.

Figure 1 Location of HTCs

3.3. What is the ABDR?

The Australian Bleeding Disorders Registry (ABDR) is a database that is designed to collect all clinical information related to the treatment of people with bleeding disorders. This includes information about patient diagnosis, including viral status, treatment details, hospital admissions and administrative information as well as details on ordering, supply and use of clotting factor products. Information is entered into the ABDR web enabled software by staff at HTCs. The current version of the ABDR has been in existence since December 2008 and background on the development of the system is at Appendix B.

3.4. ABDR Management and Governance

The ABDR is managed on a day to day basis by the NBA in accordance with the guidance and policy oversight provided by the ABDR Steering Committee. The Committee consists of representatives of the key stakeholders involved in the clinical management, advocacy and funding of treatment for people with bleeding disorders. In 2009/10 the Steering Committee representatives were:

- Dr John Rowell – Chair – representative of Australian Haemophilia Centre Directors' Organisation (AHCDO)
- Dr Chris Barnes – Chair, Australian Haemophilia Centre Directors' Organisation (AHCDO)
- Ms Sharon Caris – Executive Director, Haemophilia Foundation Australia (HFA)
- Mr Geoff Simon, Queensland Health – Jurisdictional Blood Committee nominee
- Ms Stephanie Gunn – Deputy General Manager, National Blood Authority

3.4.1. Accessing the data

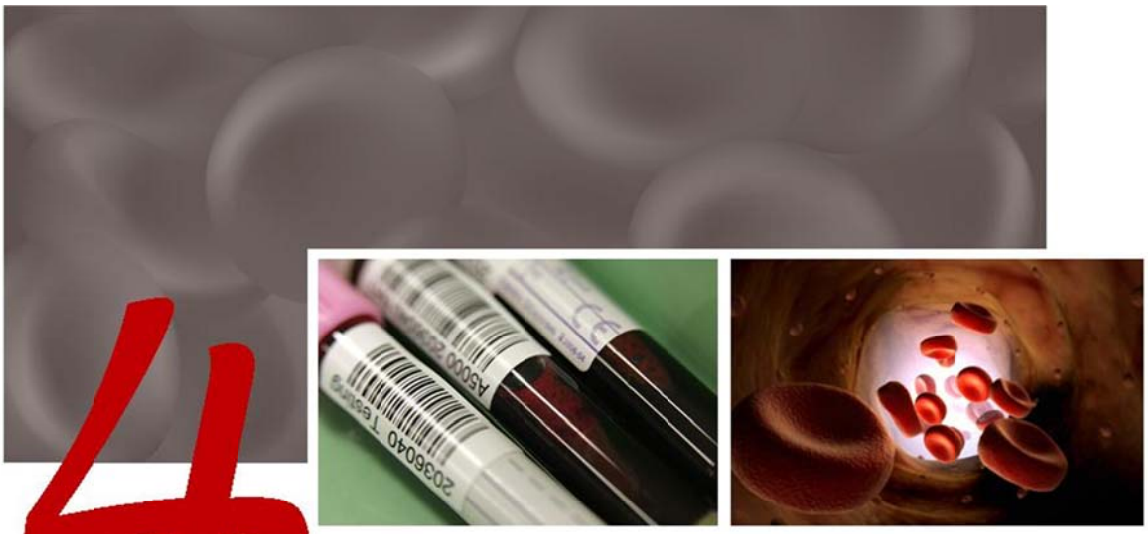
Patient confidentiality is paramount with personal data available only to the individual treating HTC and levels of authorisation/access determined by the interaction of staff with individual patients. National reporting is with aggregate, de-identified, data. All use of data, and discussion about the system, other than within the individual HTC, is considered by the ABDR Steering Committee.

The database provides a capability for all HTC staff to enter data on the interactions with patients to provide treating clinicians with a comprehensive picture of the health and wellbeing of patients. The database provides for both real time ordering of product and retrospective collection of data to provide national factor usage data to inform and assist planning and funding. Future development of the system will provide for inclusion of information on physiotherapy and social work interactions with patients.

To ensure appropriate management of the information, the NBA has instigated a detailed governance framework for a data analyst to use a Business Intelligence tool to store and access the de-identified data. This tool is called Big Red. It includes data marts for the Integrated Data Management System (IDMS – which records products issued by suppliers) and the ABDR. Big Red can provide fixed reports and *ad hoc* queries.

3.5. Data quality issues

There are a number of data quality issues in the ABDR. These include incomplete records with missing fields or "not stated" entries. As the system is still relatively young, the data in some fields is also characterised by a lack of consistency in the interpretation of specific fields. These caveats are highlighted on specific tables. The Steering Committee has initiated strategies to improve the data quality and over time the reporting from the ABDR will become more robust.



4.

Patients in the Registry

This section of the report presents the key patient data collected by the ABDR. The determination of when a person is in the register is based on their record creation date. Where another data field implied an earlier creation date the earlier date was used.

4.1. By diagnosis

Table 3 Number of people in the register and treated by latest broad diagnosis

	In register at 30 Jun 2009	Number who received product in 2008-09	In register at 30 Jun 2010	Number who received product in in 2009-10
HmA†	1836	690	1927	830
HmB†	443	152	463	185
vWD	1608	100	1748	183
Other Factor Deficiency	208	18	229	18
Platelet Disorder	145	1	154	4
Vascular	4	0	4	0
Other	35	2	40	0
Not Specified	112	2	115	0
Grand Total	**4391**	**965**	**4680**	**1220**

Note: † Includes some female carriers who are symptomatic

Table 3 confirms that Haemophilia A, von Willebrand disease and Haemophilia B are the most common disorders. Proportionally more Haemophilia A and Haemophilia B patients receive treatment. vWD is typically a mild disorder and data collected within the ABDR represents only a proportion of the total population affected with vWD. In addition, data on carriers is not complete and may not be recorded correctly.

These figures demonstrate a growth in the number of people receiving treatment with the total increasing from 965 in 2008-09 to 1220 in 2009-10.

Table 4 Number of people in the register and treated by detailed diagnosis HmA, HmB and vWD

	In register at 30 Jun 2009	Number who received product in 2008-09	In register at 30 Jun 2010	Number who received product in in 2009-10
Factor VIII Deficiency (Haemophilia A)	1581	677	1649	807
Asymptomatic Carrier Factor VIII Deficiency (Haemophilia A)†	231	7	246	14
Acquired Factor VIII Inhibitor (Acquired Haemophilia A)	24	6	32	9
Factor IX Deficiency (Haemophilia B)	388	150	401	176
Asymptomatic Carrier Factor IX Deficiency (Haemophilia B) †	55	2	62	9
von Willebrand Disease‡ - Uncharacterised	411	8	437	20
von Willebrand Disease Type 1	897	32	986	77
von Willebrand Disease Type 2 - Uncharacterised	88	7	96	18
von Willebrand Disease Type 2A	59	11	60	15
von Willebrand Disease Type 2B	39	6	42	11
von Willebrand Disease Type 2M	55	12	66	11
von Willebrand Disease Type 2N	15	2	16	2
von Willebrand Disease Type 3	37	18	38	25
Acquired von Willebrand Factor Disease	7	4	7	4
Grand Total	**3887**	**942**	**4138**	**1198**

Notes: † The practice of applying definitions does at this stage vary between HTCs and future work will focus on ensuring consistent approaches are used.

It is rare for asymptomatic carriers to require product. They do not get spontaneous haemorrhages, but may need product support in times of major trauma, surgery or at parturition.

‡ Not all with von Willebrand disease are treated through HTCs and these figures therefore do not represent the total number of vWD patients in Australia. Note also that the specific classification of vWD is incomplete in the ABDR at this stage.

Table 5 Number of people in the register and treated by diagnosis of "other disorders"

	In register at 30 Jun 2009	Number who received product in 2008-09	In register at 30 Jun 2010	Number who received product in in 2009-10
Factor V Deficiency	11	2	13	1
Factor VII Deficiency	47	6	48	6
Factor X Deficiency	9	2	10	1
Factor XI Deficiency	110	3	126	3
Factor XIII Deficiency	16	5	16	7
Fibrinogen - Afibrinogenemia	4	2	5	0
Fibrinogen - Dysfibrinogenemia	6	0	7	0
Fibrinogen - Hypofibrinogenemia	2	0	2	0
Fibrinogen Dysfunction - Uncharacterised	0	0	1	0
Vascular disorders - Ehlers Danlos Syndrome	4	0	4	0
Other (Please specify)†	112	2	115	0
Unknown†	23	0	25	0
Total	**344**	**22**	**372**	**18**

Note: † This represents incomplete data which will be addressed for the next year's report.
Those with fibrinogen disorders may have been treated with the only funded source of fibrinogen – cryoprecipitate which is not routinely collected within the ABDR.

Table 4 and Table 5 indicate that the growth in numbers of people registered in the ABDR is consistent with the historical proportion of this group of patients to the overall Australian population. This proportion and the growth is largely consistent with that illustrated in the United Kingdom Haemophilia Centre Doctors' Organisation (UKHCDO[2]) annual report. Both show a growth in the year of about 5 per cent. It is expected that with further improvements in data quality, these comparisons will be made with greater confidence.

Table 6 Number in register and receiving product by diagnosis of "Platelet disorders"

	In register at 30 Jun 2009	Number who received product in 2008-09	In register at 30 Jun 2010	Number who received product in in 2009-10
Platelet - Bernard-Soulier	1	0	1	0
Platelet - Glanzmann's Thrombasthenia	6	0	7	2
Platelet - May Hegglin	3	0	3	0
Platelet - Storage Pool (Dense Granule) Deficiency	7	0	11	0
Platelet – Uncharacterised†	128	1	132	2
Total	**145**	**1**	**154**	**4**

Notes: † This represents incomplete data which will be addressed for the next year's report.
Platelet disorders may be treated with DDAVP, platelet infusion (not routinely collected in the ABDR) or rVIIa (collected within the ABDR).

[2] http://www.ukhcdo.org/

Table 6 shows that data on platelet disorders is not heavily recorded in the ABDR. These disorders tend to be under reported and are typically treated at facilities outside the HTC framework and will, therefore, not necessarily be recorded in the ABDR. Nevertheless, this field will be more value when clear definitions are applied to those patients currently recorded in the ABDR as uncharacterised and this will be a priority for future work.

Table 7 People in the register at 30 Jun 2010 by broad age group, diagnosis and severity

	Paediatric 0–19 years		Adult 20 years and over		Total	
	In register at 30 Jun 2010	**Number receiving product in 2009-10**	**In register at 30 Jun 2010**	**Number receiving product in 2009-10**	**In register at 30 Jun 2010**	**Number receiving product in 2009-10**
HmA†	**550**	**330**	**1377**	**500**	**1927**	**830**
Severe	262	232	322	257	584	489
Moderate	76	48	172	80	248	128
Mild	204	49	776	153	980	202
Not Applicable‡			19	2	19	2
Unknown	8	1	88	8	96	9
HmB†	**120**	**63**	**343**	**122**	**463**	**185**
Severe	43	37	45	35	88	72
Moderate	27	17	74	35	101	52
Mild	47	8	206	52	253	60
Not Applicable‡			3	0	3	0
Unknown	3	1	15	0	18	1
vWD	**407**	**38**	**1341**	**145**	**1748**	**183**
Grand Total	**1077**	**431**	**3061**	**767**	**4138**	**1198**

Note: Mild cases of HmA, HmB and vWD are often treated with DDAVP and the use of this product may not be recorded in the ADBR.

† Includes some female carriers.

‡ Not applicable – may represent data input inconsistencies or incomplete data, and will be addressed for next year's report.

Unknown represent patients not yet classified or symptomatic carriers not classified.

Table 7 illustrates that a higher proportion of paediatric patients with HmA receive product than do adults and that this difference is less marked for patients with HmB.

Figure 2 Age distribution of people in the ABDR by diagnosis at 30 June 2010

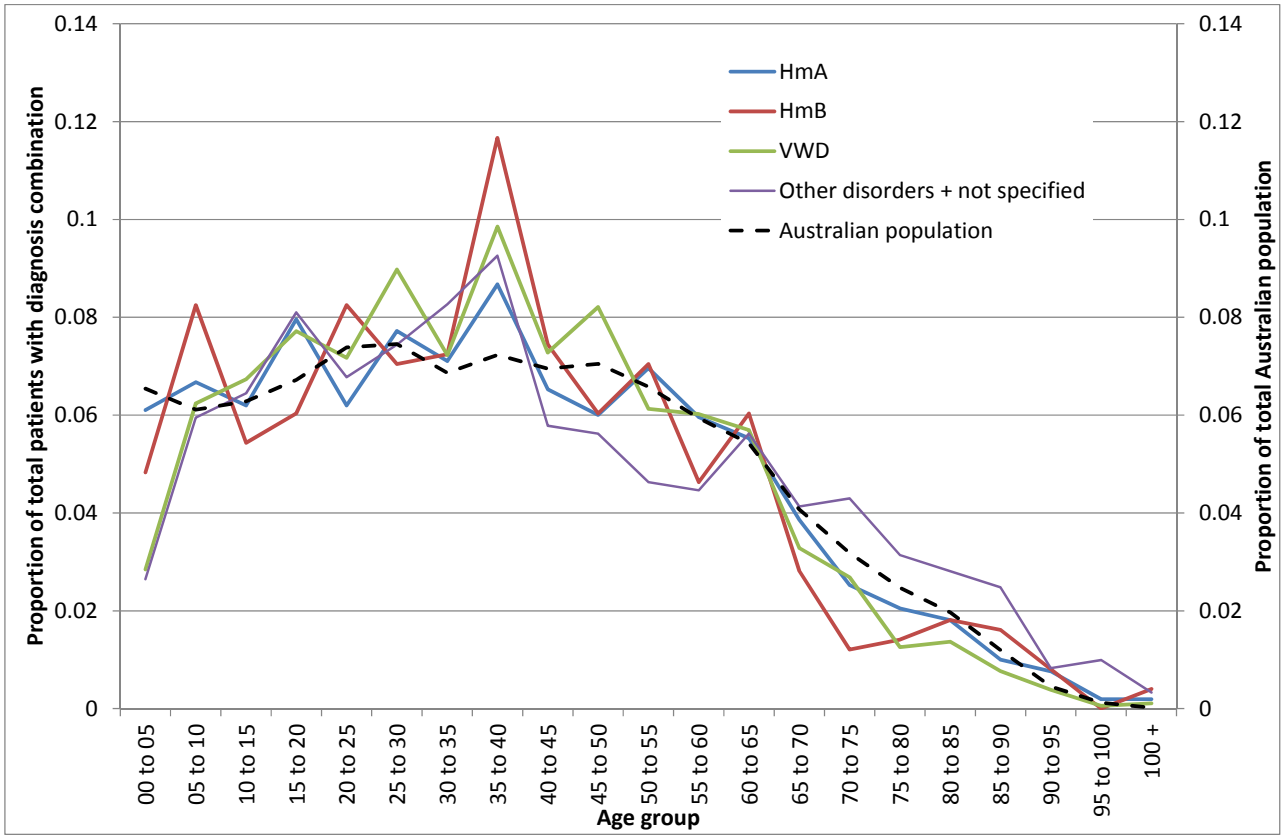


Figure 2 illustrates that the proportion of patients with all disorders currently aged between 35-40 is higher than the proportion of the Australian population at that age group. Notably the proportion of patients aged 5-10 with HmB and those aged between 15-20 with HmA are also substantially higher than the proportion of the Australian population at that age. However, the proportion of patients aged between 60 and 80 with all traditional forms of bleeding disorders is substantially lower than that for the rest of the population. The proportion with other bleeding disorders which are more typically associated with aging and associated medical treatments is the reverse of this picture.

Table 8 von Willebrand Disease in the register at 30 Jun 2010 by broad age group and vWD classification

	Paediatric 0–19 years		Adult 20 years and over		Total	
	In register at 30 Jun 2010	Number receiving product 2009-10	In register at 30 Jun 2010	Number receiving product 2009-10	In register at 30 Jun 2010	Number receiving product 2009-10
von Willebrand Disease Type 1	214	9	772	68	986	77
von Willebrand Disease Type 2A	10	0	50	15	60	15
von Willebrand Disease Type 2B	9	2	33	9	42	11
von Willebrand Disease Type 2 - Uncharacterised	34	7	62	11	96	18
von Willebrand Disease Type 2M	24	5	42	6	66	11
von Willebrand Disease Type 2N			16	2	16	2
von Willebrand Disease Type 3	10	5	28	20	38	25
Acquired von Willebrand Factor Disease			7	4	7	4
von Willebrand Disease - Uncharacterised	106	10	331	10	437	20
Grand Total	407	38	1341	145	1748	183

While vWD is typically a mild disorder and data collected within the ABDR represents only a proportion of the total population affected with vWD. This data will be interesting to track over time as there is now an increased awareness of this disease and it is often diagnosed during surgical interventions. As the population ages and seeks more medical intervention, this disease may become more heavily reported. If they receive plasma derived von Willebrand factor they must be included in the ABDR.

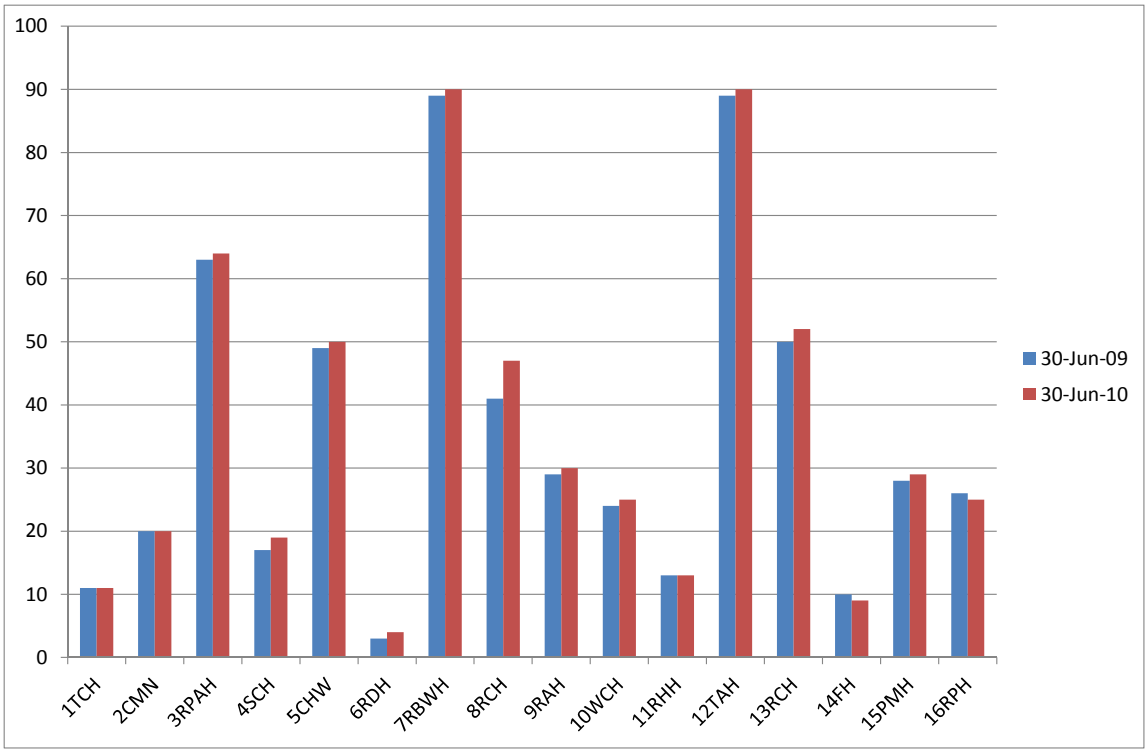
Table 9 Comparison of the proportion of patients in the register and treated, UK and Australia, major diagnoses 2010

	Female			Male		
	In Register 2009-10	Treated 2009-10	Proportion treated	In Register 2009-10	Treated 2009-10	Proportion treated
Australia						
HmA (Symp + Asymp)	294	25	8.5%	1631	805	49.4%
HmB (Symp + Asymp)	80	15	18.8%	383	170	44.4%
vWD	1003	96	9.6%	745	87	11.7%
Other conditions	278	9	3.2%	264	13	4.9%
UK						
HmA (Symp + Asymp)	1082	53	4.9%	5346	2863	53.6%
HmB (Symp + Asymp)	332	37	11.1%	1125	618	54.9%
vWD[3]	5588	620	11.1%	3268	363	11.1%

The UK population is approximately three times that of Australia. The treatment rates shown in Table 9 are comparable between Australia and the UK although on current data, the proportion of Australian HmB patients receiving product does appear to lower. The incidence of most conditions seems higher in the UK than Australia.

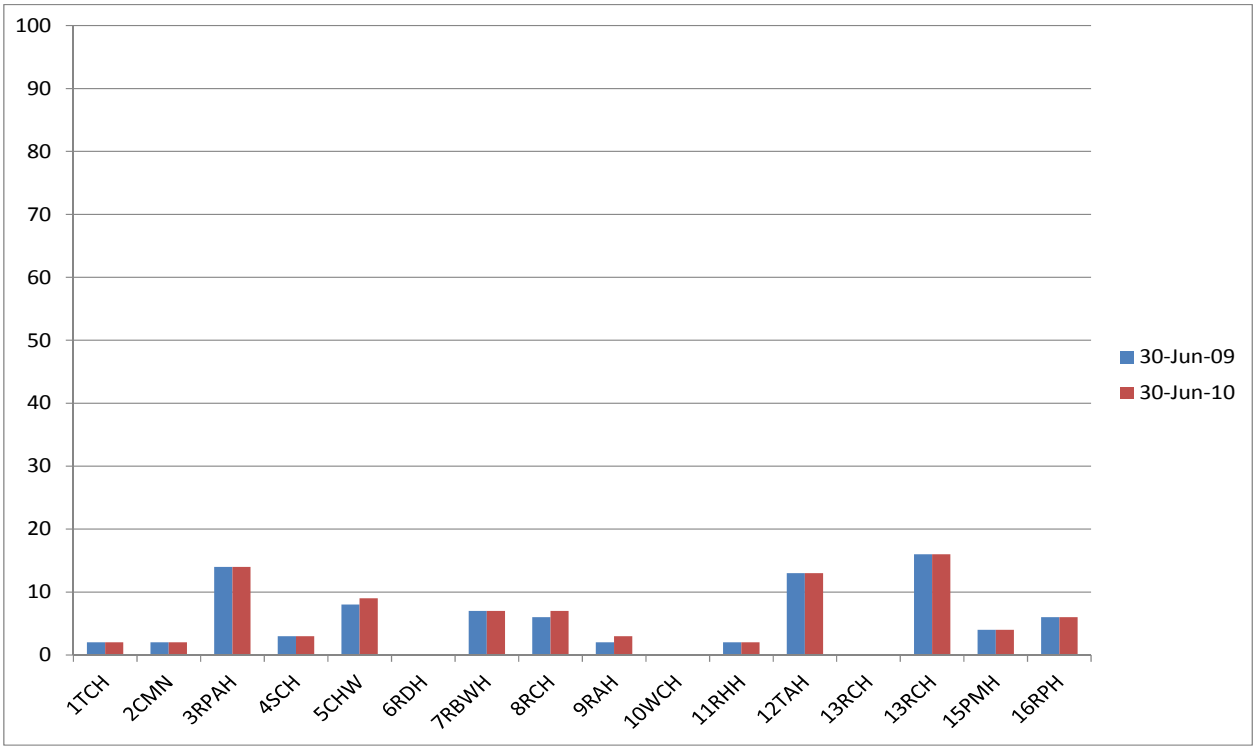
4.2. By geography

Figure 3 Distribution of severe Haemophilia A patients by HTC by year



[3] The UK publication did not have a split of treated by gender so we have split the treated in proportion to number in register.

Figure 4 Distribution of severe Haemophilia B patients by HTC by year



The figures above illustrate variability in the numbers of severe patients registered in each HTC mainly reflecting their catchment size and modest growth in numbers of severe patients numbers over time.

Figure 5 People in the register at 30 June 2010 with Haemophilia A or Haemophilia B by severity and home state (grouping small states)

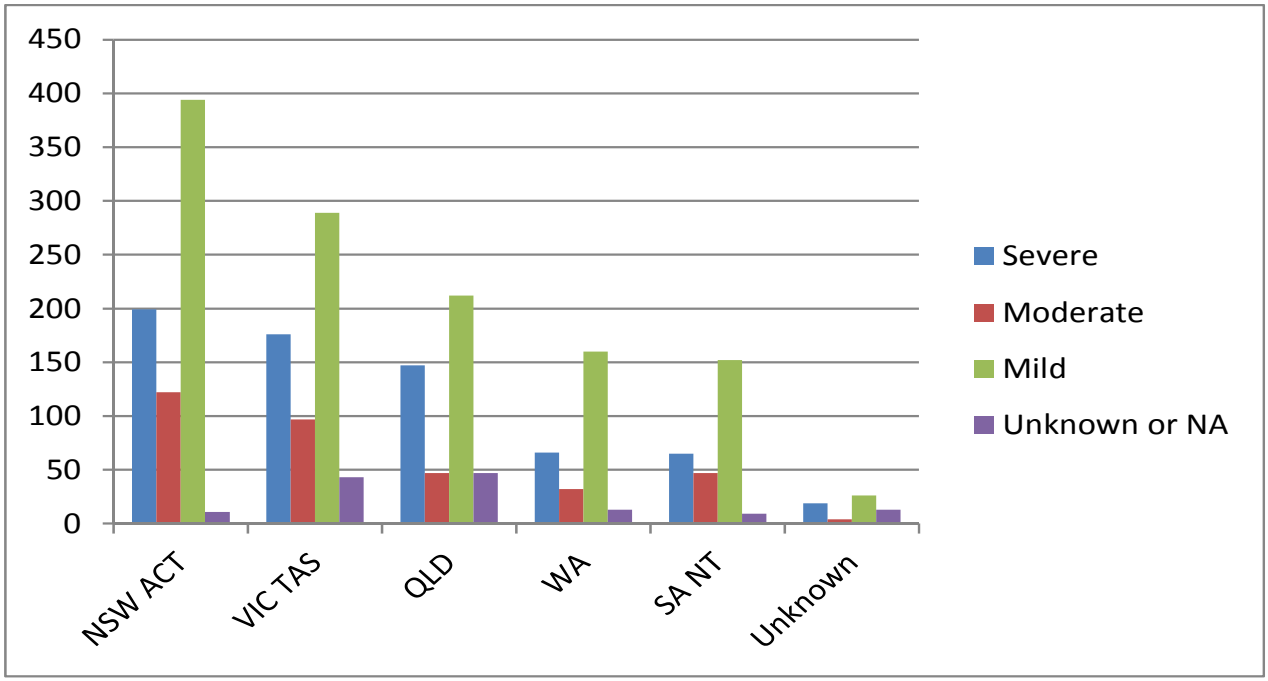


Figure 5 illustrates that numbers of patients with haemophilia are largely consistent with population size. Unknown includes patients who may be temporally overseas and patients who have less severe conditions and have not had recent contact with their HTC.

4.3. Incidence of major disorders

Table 10 shows that Haemophilia A in males has the highest incidence with nearly 15 per 100,000 males. The next highest incidence is for von Willebrand disease in females at nearly 9 per 100,000.

Table 10 Incidence of major disorders in Australia people with haemophilia per 100,000 of relevant population

	Female		Male		Persons	
	In register at 30 Jun 2009	In register at 30 Jun 2010	In register at 30 Jun 2009	In register at 30 Jun 2010	In register at 30 Jun 2009	In register at 30 Jun 2010
HmA all	2.4	2.6	14.4	14.7	8.4	8.6
HmA Severe	0.1	0.1	5.1	5.1	2.6	2.6
HmB all	0.6	0.7	3.4	3.4	2.0	2.1
vWD all	8.3	8.9	6.3	6.7	7.3	7.8

The numbers in Table 10 are calculated by dividing the number of people in the register at the date with the broad diagnoses by the total corresponding Australian estimates population at the same date and multiplying by 100,000.

Table 11 Incidence of bleeding disorders selected countries 2008 (per 100,000)

Country	Population	B1. Number of PWH[4] (HmA or HmB)	B2. Number of people with vWD	B3. Number of people with OBD[5]	Number of PWH per 100,000	Number of people with vWD per 100,000	Number of people with OBD per 100,000
Australia	**21,262,641**	**1,760**	**1,308**	**629**	**8.3**	**6.2**	**3.0**
France	64,057,792	4,779	272	215	7.5	0.4	0.3
Germany	82,329,758	4,000	745		4.9	0.9	0.0
Netherlands	16,715,999	1,452	263	64	8.7	1.6	0.4
Spain	40,525,002	1,932	690	199	4.8	1.7	0.5
Sweden	9,059,651	1,017	1,523		11.2	16.8	0.0
United Kingdom	61,113,205	6,061	7,852	4,999	9.9	12.8	8.2
United States	307,212,123	16,243	11,852	1,616	5.3	3.9	0.5

Source: World Federation Hemophilia Report on the annual global survey 2008

[4] PWH means people with haemophilia.

[5] OBD means other bleeding disorders.

Table 12 Incidence of Haemophilia A in males in OECD countries (per 100,000)

Country	1998	1999	2000	2001	2002	2003	2004	2005	2006	Mean
Iceland	37.7	NA	NA	39.4	NA	39.3	38.1	38.5	NA	38.6
United Kingdom	19.0	19.4	17.4	17.6	NA	17.2	NA	22.6	20.7	19.1
Netherlands	15.3	16.0	15.9	17.7	NA	17.5	18.6	18.0	18.5	17.2
New Zealand	18.5	17.0	16.7	21.6	NA	17.8	11.7	12.3	21.9	17.2
Ireland	12.5	16.5	17.5	16.7	NA	16.6	17.8	18.9	18.3	16.8
Sweden	15.5	16.2	14.9	NA	NA	14.9	15.0	NA	NA	15.3
Switzerland	14.5	13.3	14.6	11.9	NA	12.1	13.2	12.7	14.2	13.3
Canada	11.9	13.1	12.6	12.5	12.4	NA	14.0	14.3	14.6	13.2
Denmark	11.8	11.8	12.5	12.4	NA	12.6	12.1	13.1	NA	12.3
Czech Republic	12.2	12.2	12.2	NA	NA	NA	NA	NA	NA	12.2
France	12.7	12.7	14.8	NA	NA	NA	NA	9.8	11.0	12.2
Greece	10.7	10.8	12.2	12.4	NA	12.4	12.8	13.0	13.0	12.2
Belgium	10.5	11.8	12.1	12.4	12.4	12.3	NA	NA	NA	11.9
Norway	11.6	11.5	11.6	NA	NA	11.9	NA	12.3	12.2	11.9
Italy	9.0	12.3	NA	NA	NA	NA	13.8	13.8	9.4	11.7
Australia	**10.8**	**10.6**	**10.5**	**10.4**	**NA**	**NA**	**8.8**	**12.8**	**13.5**	**11.1**
Luxembourg	10.0	9.0	13.5	NA	NA	NA	NA	NA	NA	10.8
Germany	NA	13.2	9.6	9.8	NA	10.0	10.0	10.0	10.0	10.4
Portugal	8.7	8.7	8.7	8.6	NA	10.3	9.6	9.6	9.7	9.2
Finland	NA	NA	NA	NA	NA	8.6	NA	9.0	9.1	8.9
Spain	10.9	8.5	8.5	10.0	NA	9.7	7.6	7.6	7.6	8.8
Austria	NA	8.4	8.4	NA	NA	8.7	8.7	NA	NA	8.5
United States	7.6	7.5	7.7	7.8	NA	7.6	7.8	7.9	8.0	7.8
Japan	5.3	5.5	NA	5.9	NA	6.2	NA	6.3	6.5	5.9
Korea	4.7	5.0	5.2	5.3	5.3	NA	5.5	5.8	5.9	5.3

Source: World Federation of Hemophilia – Facts and figures (December 2010)

Table 12 shows that of the OECD countries Australia is placed in the middle for incidence of Haemophilia A.

Table 13 Incidence of Haemophilia A in males in selected countries by severity (per 100,000)

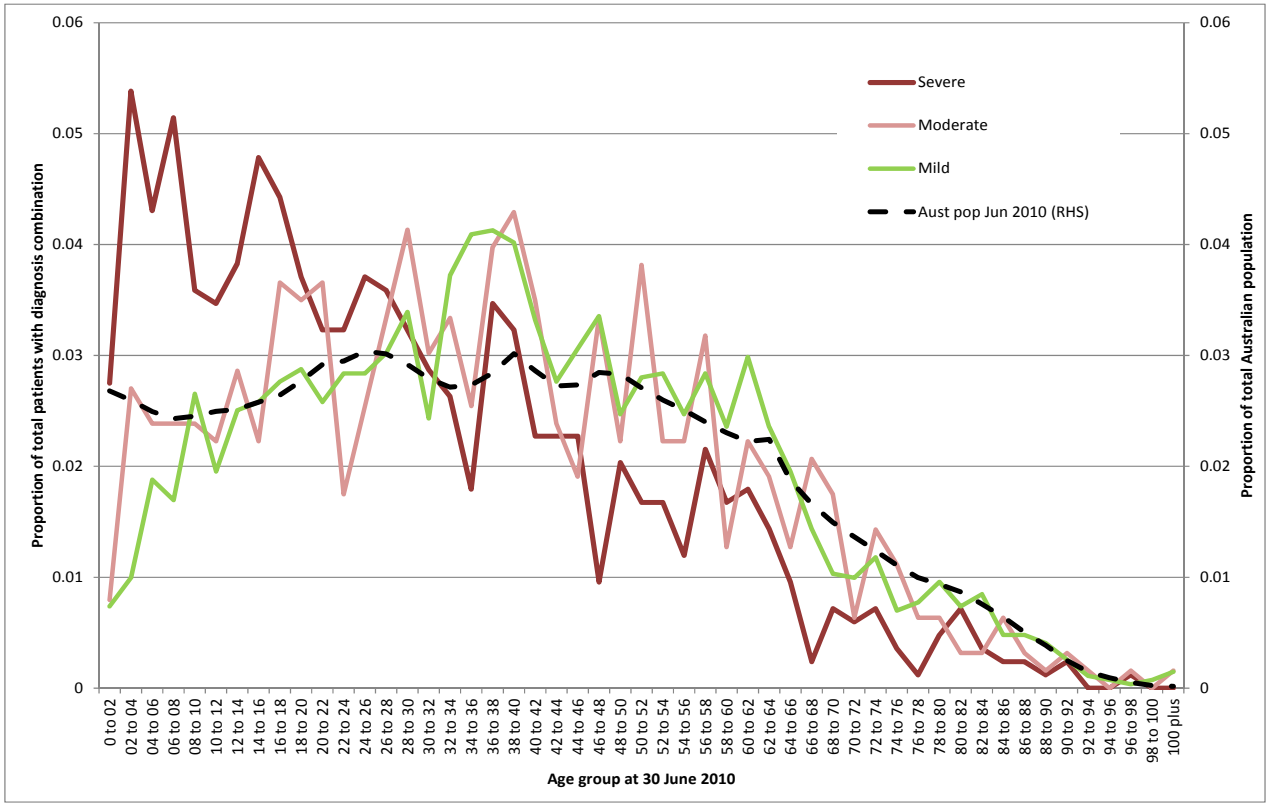
Country	Year	Overall	Severe	Severe as a proportion of overall
Canada	2008	14.3	4.3	30%
Greece	1992	12.0	3.6	30%
Italy	2006	9.5	4.8	51%
Netherlands	2001	11.7	NA	NA
United Kingdom	2008	21.5	7.1	33%
United States	1998	10.4	4.4	42%
Australia – In register (current ABDR at 30 June)	2010	14.7	5.1	35%

Source: World Federation of Hemophilia – Facts and figures (December 2010); ABDR

Table 13 shows that proportion of severe cases in Australia is broadly comparable to other countries

4.4. By age distribution

Figure 6 Age distribution of people in the register at 30 June 2010 by severity



In Figure 6 we see that the age distribution of the severe sufferers is much younger than the other groups and the Australian population as a whole. There is also a dip in the distribution of the severe group around the ages between 40 and 50. The higher proportion at younger ages reflects early diagnosis of the more severely affected.

Figure 7 Age distribution of people in the register at 30 June 2010 with Haemophilia A and Haemophilia B by severity

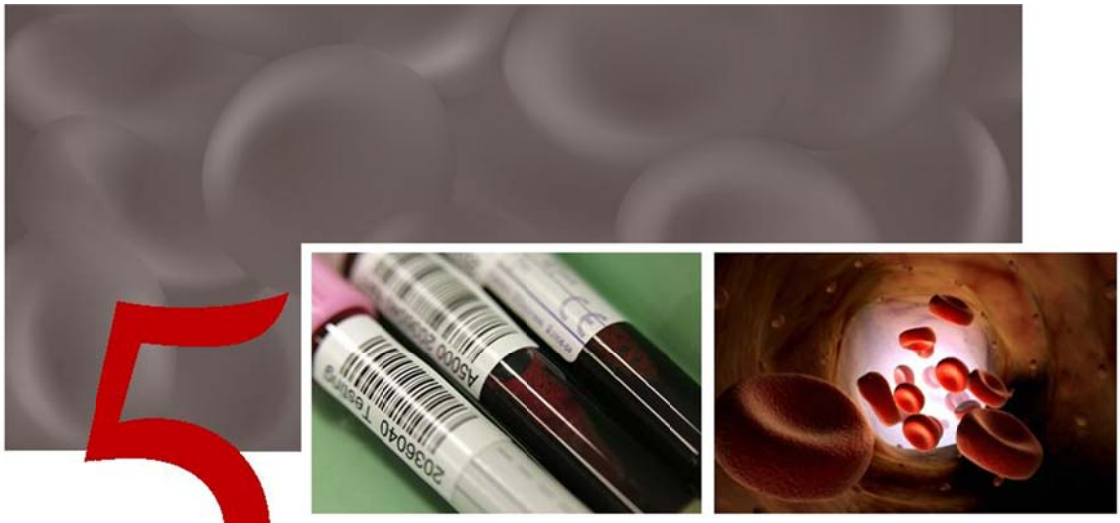
4.5. By Inhibitors

Table 14 Number of patients with inhibitors and comparison with UK

	Severe			Moderate			Mild			Total	
	Inhibitor present	proportion of total with inhibitors	Total Severe	Inhibitor present	proportion of total with inhibitors	Total Moderate	Inhibitor present	proportion of total with inhibitors	Total Mild	proportion of total with inhibitors	Total in Register at 2010
Australia											
HmA (Symp + Asymp)	111	19.0%	584	14	5.6%	248	25	2.6%	980	7.8%	1927
HmB (Symp + Asymp)	6	6.8%	88	0	0.0%	101	0	0.0%	253	1.3%	463
vWD	2	1.7%	119	0	0.0%	225	0	0.0%	1184	0.2%	1748
UK											
HmA (Symp + Asymp)	351	19.3%	1814	43	7.7%	559	62	2.1%	2972	8.5%	5345
HmB (Symp + Asymp)	15	3.8%	396	0	0.0%	244	0	0.0%	482	1.3%	1122
vWD	2	1.6%	125	2	1.2%	164	0	0.0%	8381	0.0%	8670

Inhibitors are made by the body's immune system to react against replacement clotting factor proteins. This occurs when the immune system perceives the proteins as foreign or harmful to the body. When this happens, the inhibitors prevent the replacement factors from working properly to stop bleeding.

There is some concern at the data entry consistency in this field, however the Australian data appears broadly comparable with the UK data.



5.

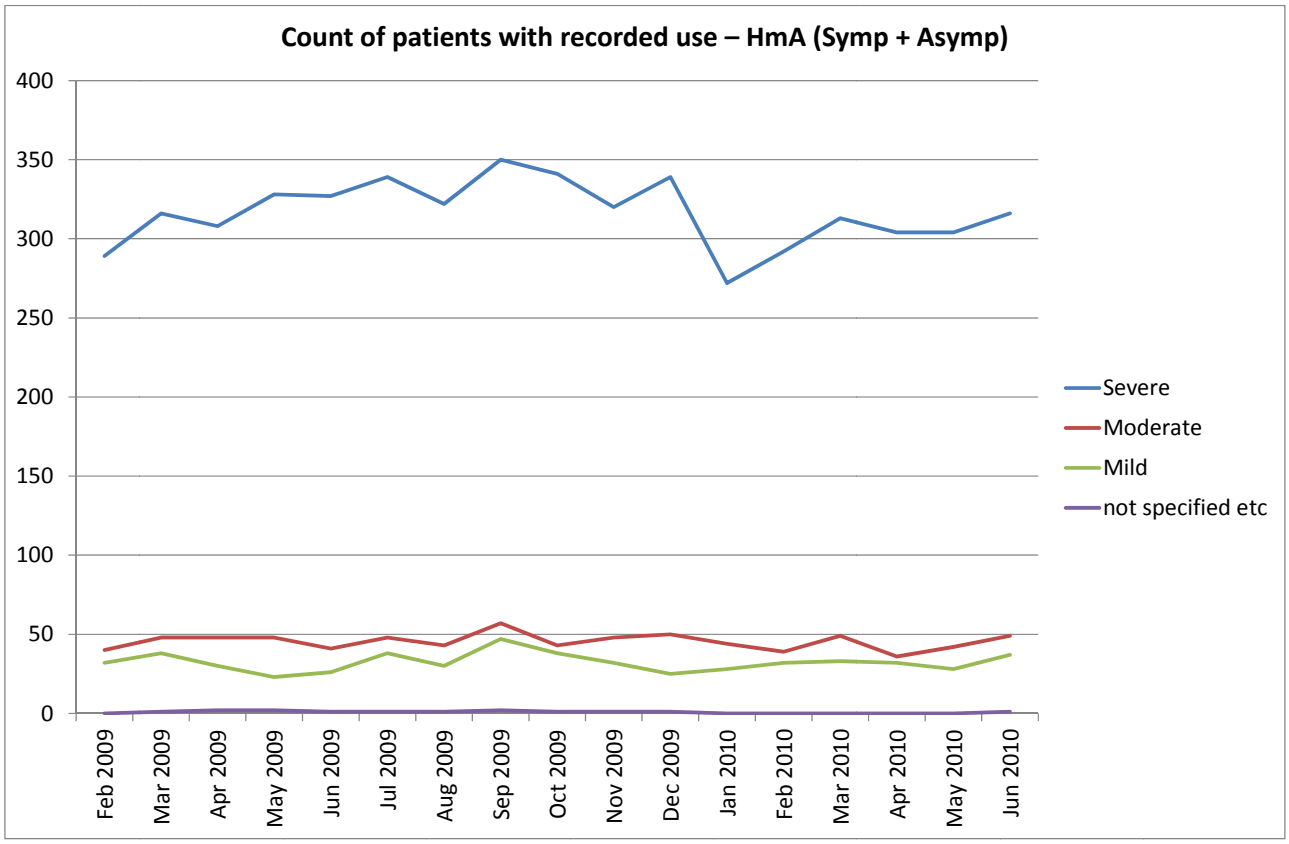
Patients who received treatment

The charts in this are based on the reports of product use in the ABDR and have been collated using the NBA's Big Red data marts.

5.1. By severity

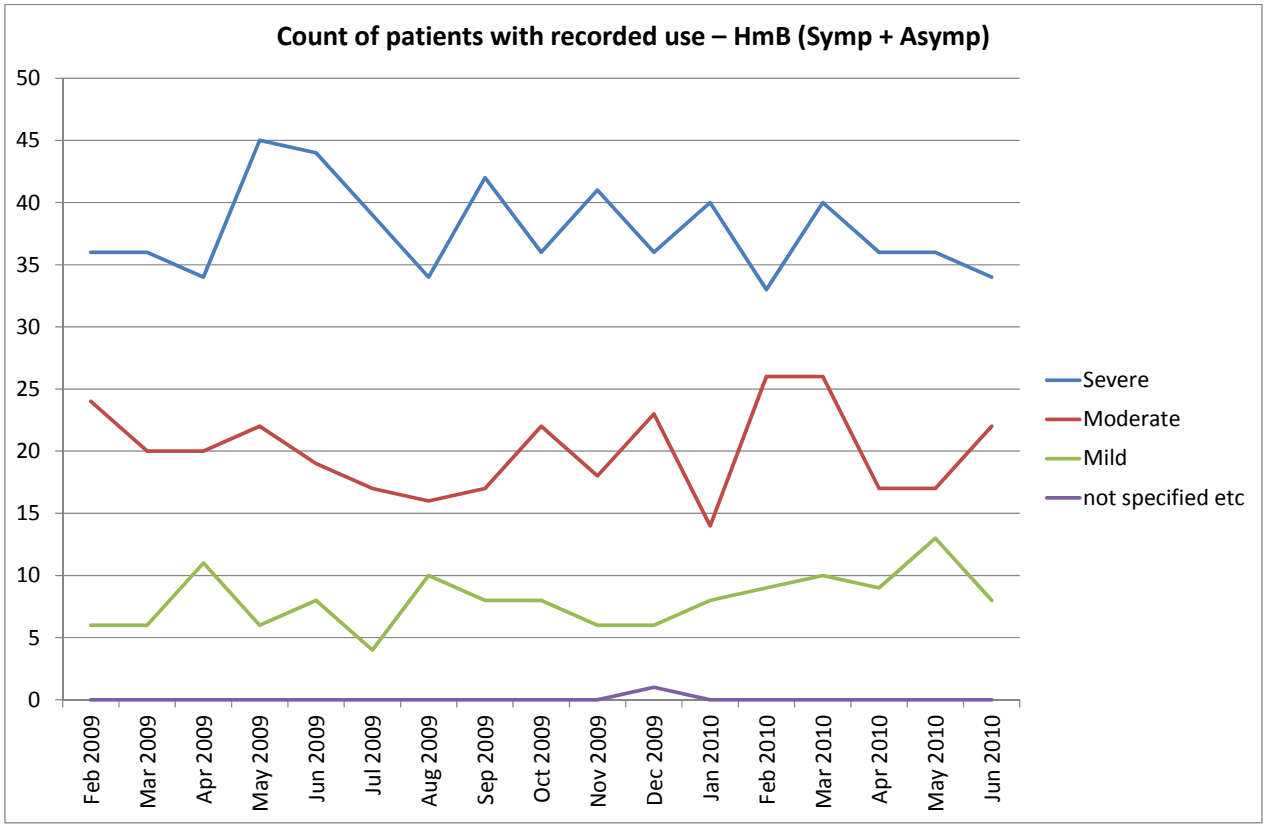
As would be expected, more severe patients are treated more often and receive more product. It is important for supply planning purposes to understand trends in the proportion of patients diagnosed as severe to ensure adequacy of supply and likely demand in the future.

Figure 8 HmA (Symp + Asymp) – Number of patients receiving treatment by severity



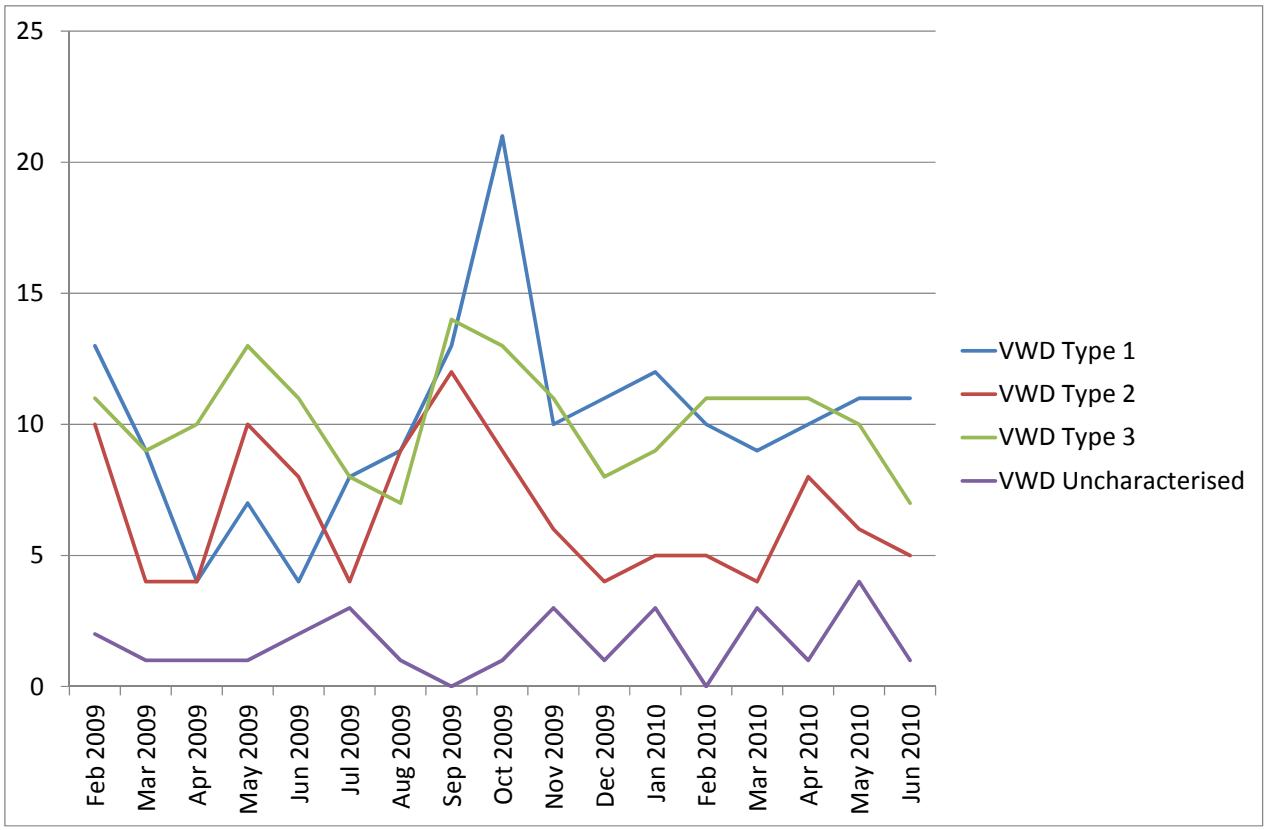
Source: ABDR

Figure 9 HmB (Symp + Asymp) – Number of patients receiving treatment by severity



Source: ABDR

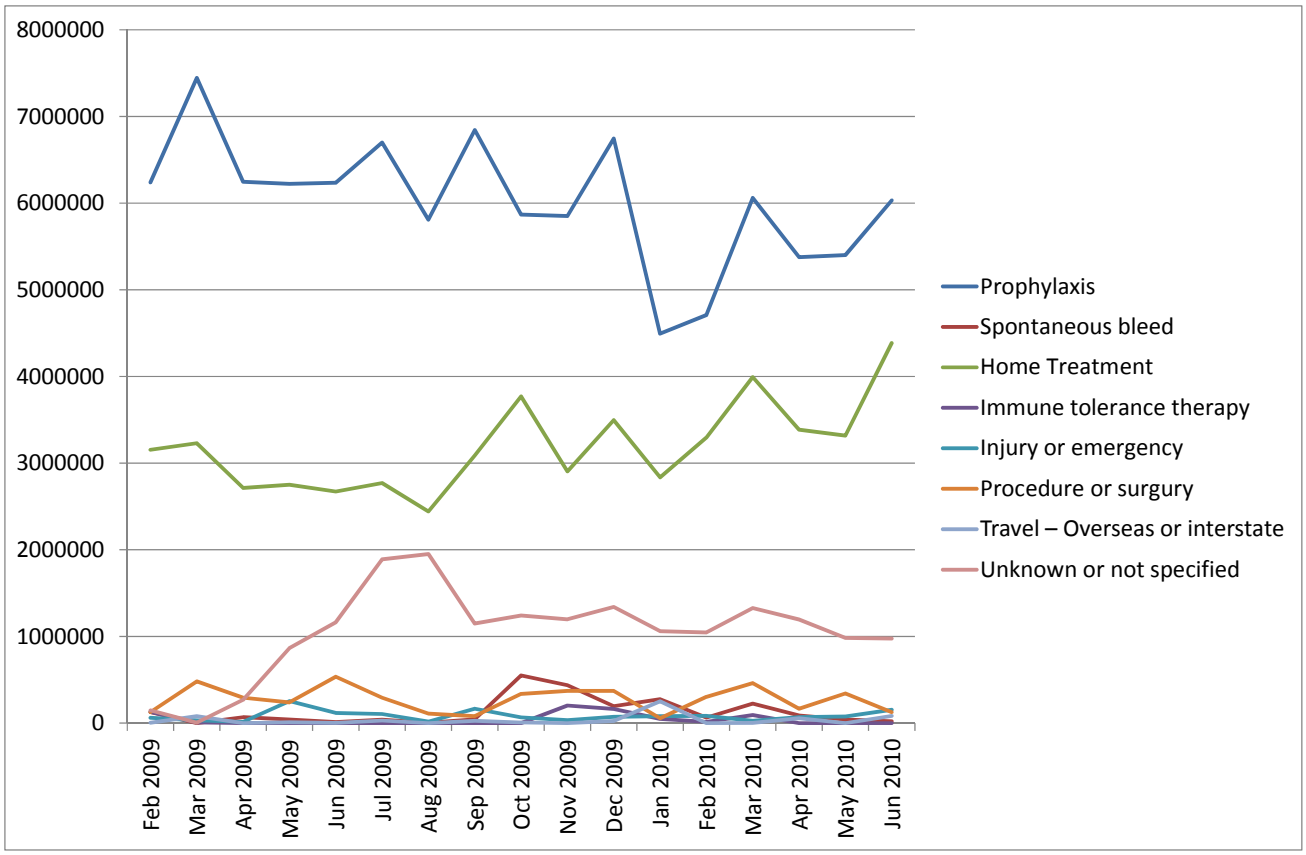
Figure 10 vWD – Number of patients receiving treatment by type of vWD



Source: ABDR

As would be expected, a higher proportion of those patients with severe diagnosis, receive treatment during the year although the pattern for patients with vWD is less predictable than other diagnoses as numbers are small. Note that many persons with vWD particularly type 1 will receive DDAVP – use of which is not recorded in the ABDR.

Figure 11 International Units of product received by patients treated by purpose



Source: ABDR

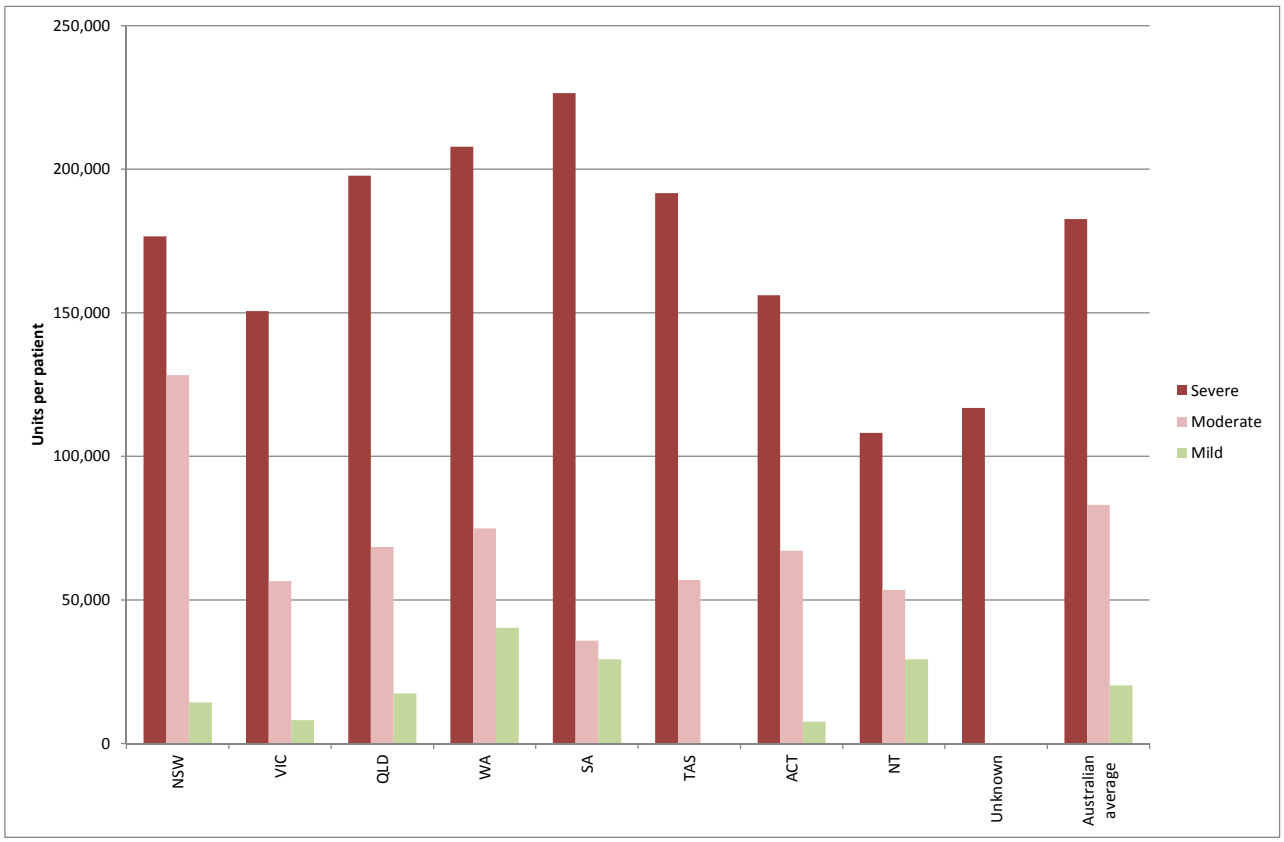
This field in the ABDR has been identified as requiring further clarification. The classifications by purpose currently provided are not mutually exclusive and some patients may have been counted more than once. Further, different HTCs or clinicians may interpret them differently. The definitions and data entry protocols will be an area of focus for future work. A likely revamp of this field will be to record this information by:

- Location – home or hospital
- Regimen – prophylaxis or on demand
- Purpose – bleed, surgery, procedure or prevention (prophylaxis)

5.2. National average issues of Factor VIII by severity

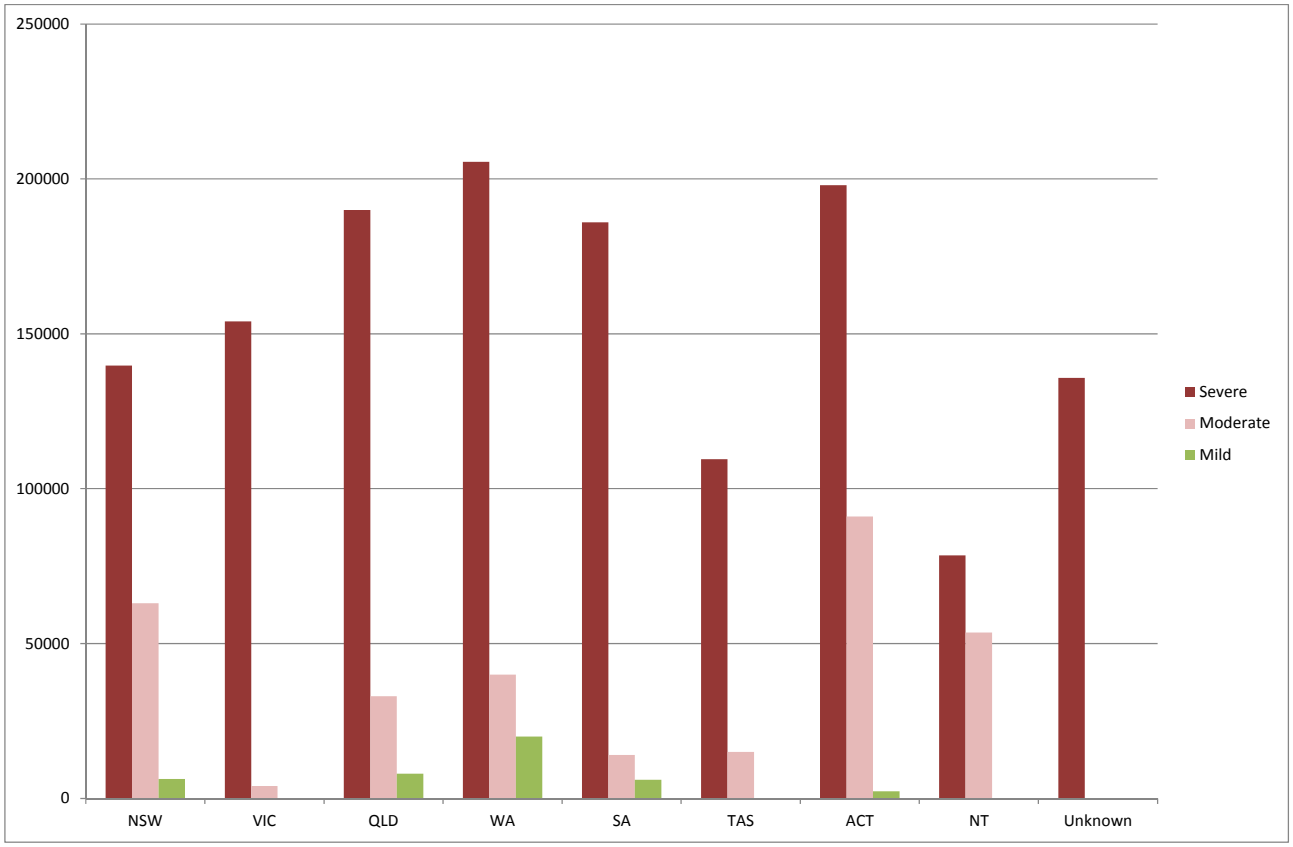
There are many other factors affecting the use of product including body weight and ranges of severity. Figure 12 shows the average issue of factor VIII by severity by state. The more severe the condition is the greater the amount of product given. Moreover, there is considerable difference between the states. Some of this difference relates, however, to incomplete record of product use at the time of publication and recording use with a not stated product.

Figure 12 Total issues of Factor VIII recorded in the ABDR issued to Haemophilia A patients divided by the number in the register who were treated, by home state of patient and severity in 2009-10



Unknown: Where location, as at the time of reporting, was indeterminate

Figure 13 Median Factor VIII per Haemophilia A patients by state and severity in 2009-10



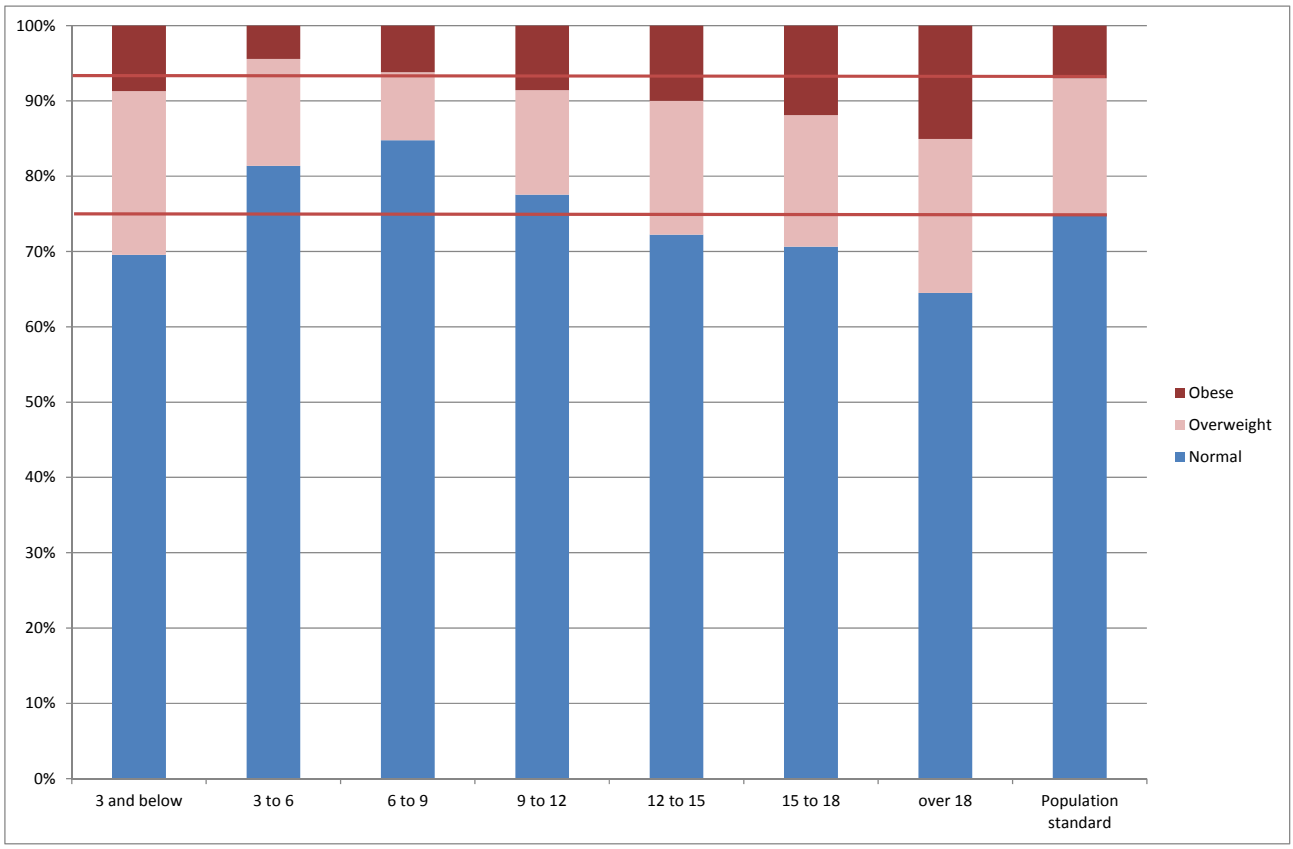
Note: Product that is not fully described is coded as zero which is the reason for some of the zero value for Mild.
Unknown: Where location, as at the time of reporting, was indeterminate

Table 15 Issues of Factor VIII to Haemophilia A patients UK and Australia

	United Kingdom			Australia		
	Number treated	Total units of FVIII	Units per patient	Number treated	Total units of FVIII	Units per patient
Severe HmA	1,596	366,276,171	229,496	489	89,335,750	182,691
Non severe HmA	1,111	64,812,779	58,337	341	14,940,250	43,813
Tot HmA	2,707	431,088,950	159,250	830	104,276,000	125,634

5.3. By weight and height

Figure 14 Weight and height data for ABDR patients receiving treatment



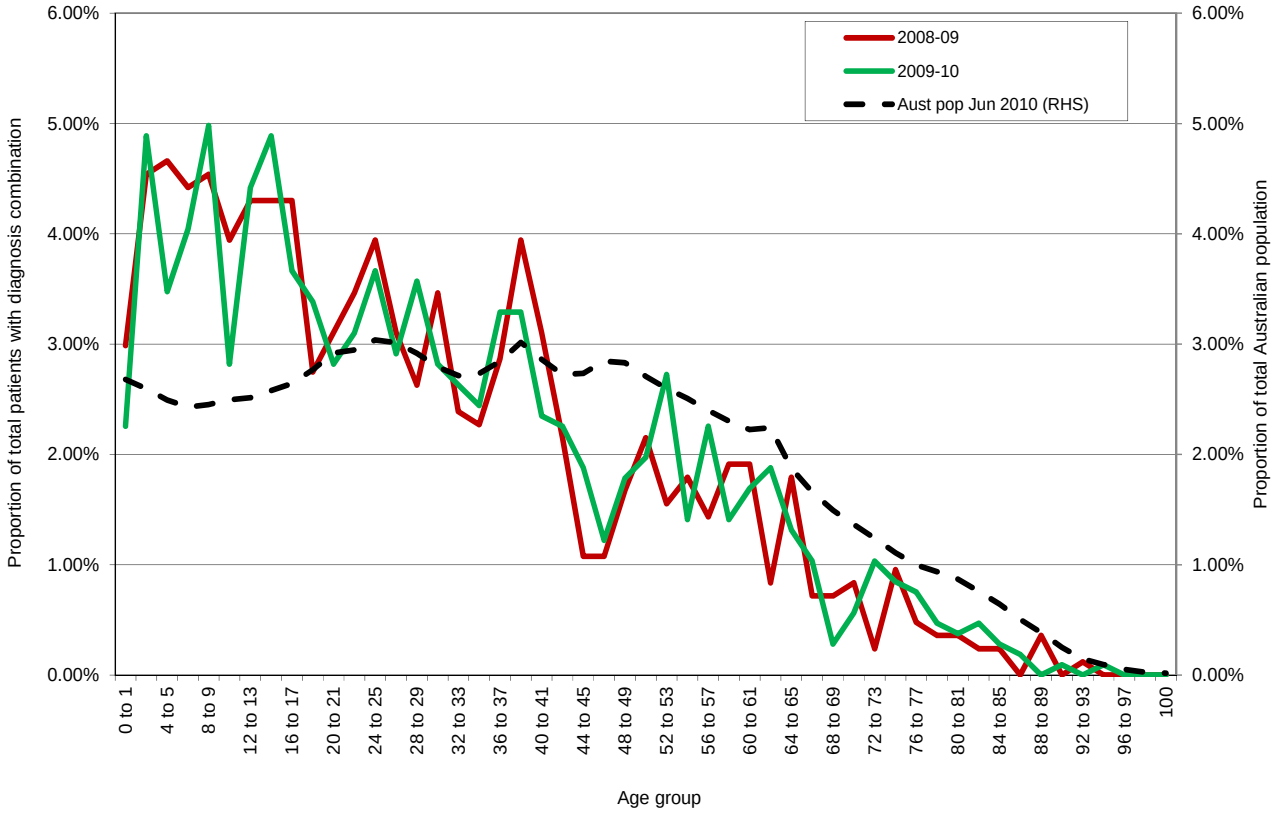
Source: ABDR and Victorian Department of Health

Figure 14 was constructed using age, weight and height data collected in ABDR. These data were combined to calculate the body mass index (BMI). The calculated BMIs were compared with an age appropriate BMI scale produced by Victorian Department of Health to determine whether the BMI corresponded to that person being overweight or obese. Generally 25 per cent of the population are considered overweight and 7 per cent obese. These data may not be fully representative for adult patients as the height and weight data is less complete for them. Nevertheless, as dosage is often related to weight high proportions of overweight and obese patients may lead to an increase in the rate of growth of demand for products.

5.4. By age distribution

The following section shows the age distribution of patients who received product in at least one of the last two financial years.

Figure 15 Age distribution of patients who received treatment in 2008-09 or 2009-10 compared with the Australian population



Source: ABDR and ABS Population statistics

Figure 15 shows that those patients in the ABDR who are receiving treatment are generally younger than the Australian population as a whole. Also of note is the dip in the distribution at patients aged around 40 to 45. These distributions are similar to that for patients with severe conditions in Figure 6 and Figure 7.

Figure 16 Age distribution at 30 June 2010 of Haemophilia A and Haemophilia B patients who received product in 2009-10

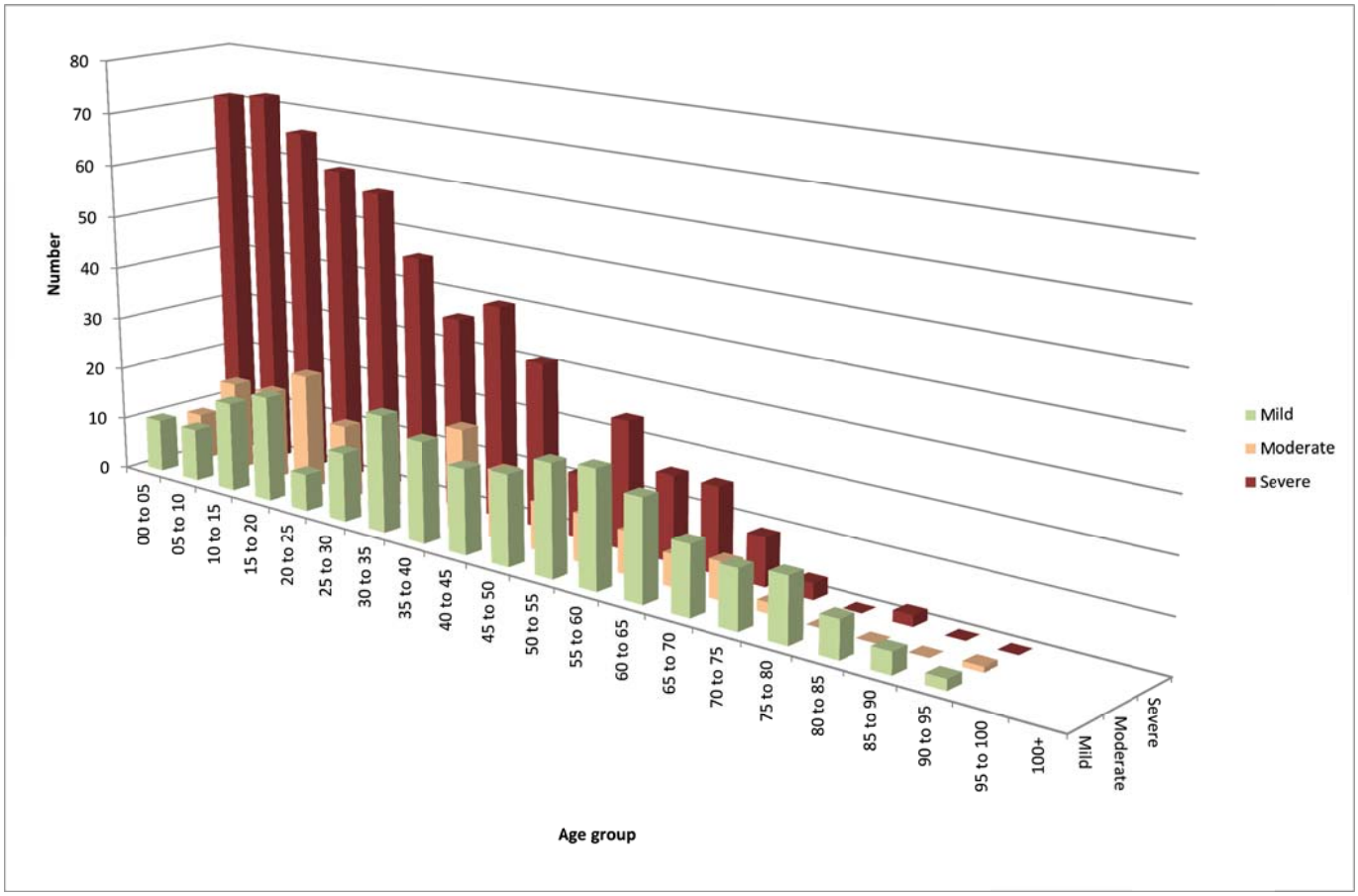
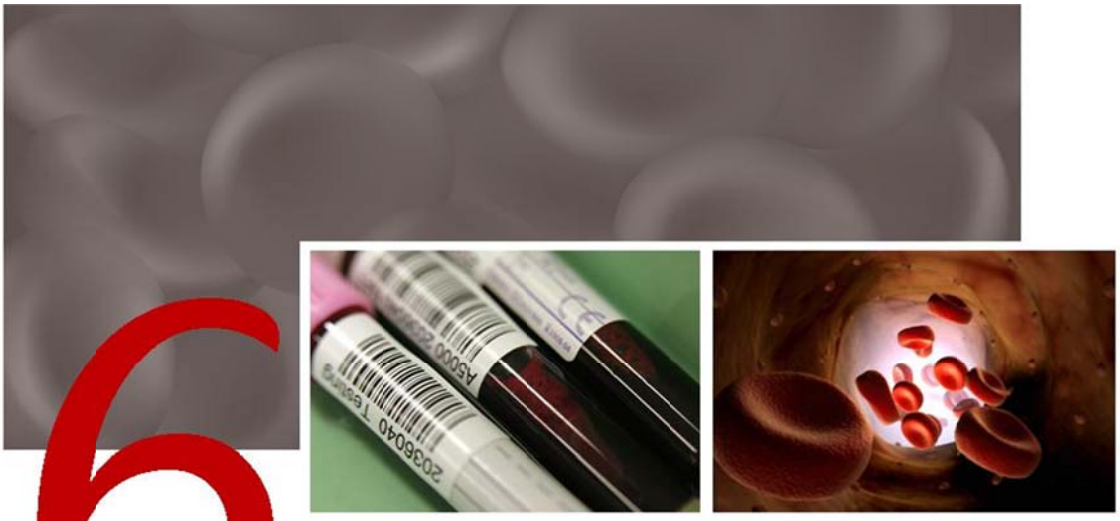


Figure 16 shows a large proportion of the severe Haemophilia A and Haemophilia B population are in the younger age groups. If the higher numbers in the younger age groups flow into older age groups and are replaced, overall demand may grow strongly.



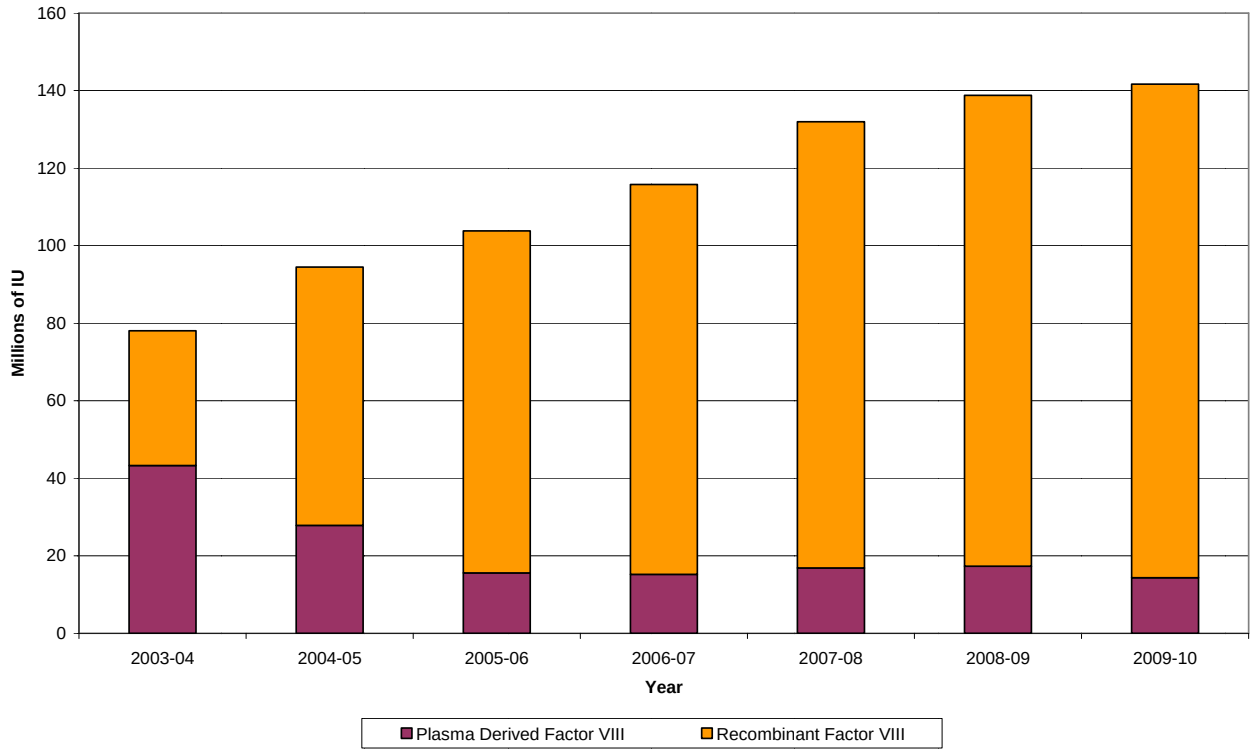
6.

Volume & cost of products used in the treatment of bleeding disorders

6.1. National issues by product

Figure 17 shows both the strong growth of clotting factors used in the treatment of haemophilia sufferers and the even stronger growth in the use of recombinant clotting factors.

Figure 17 Annual international units of Factor VIII issued nationally 2003-04 to 2009-10

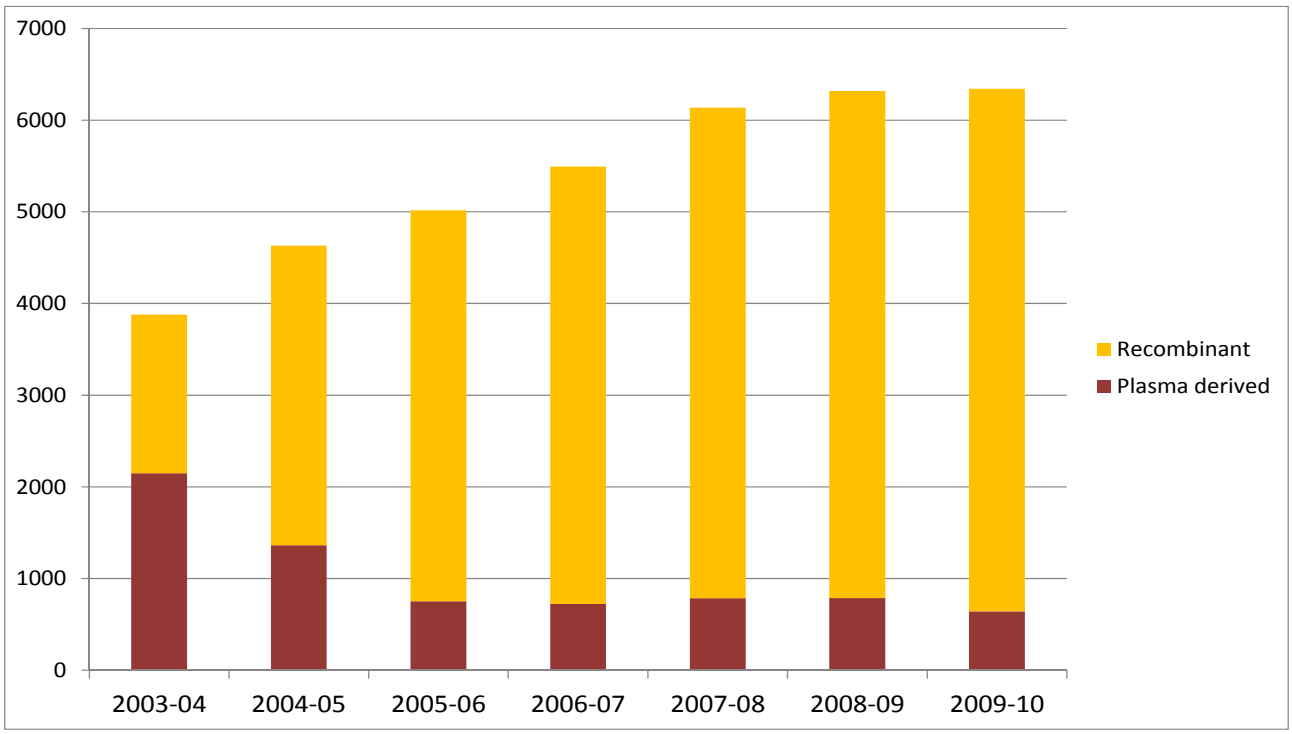


Source: IDMS database of issues

This shows that demand for Factor VIII has increased by 82 per cent over the years (an average rate of 10.4 per cent per annum) but that this growth rate in the last two years has slowed to 2.1 per cent. This figure also illustrates the rapid uptake of recombinant product.

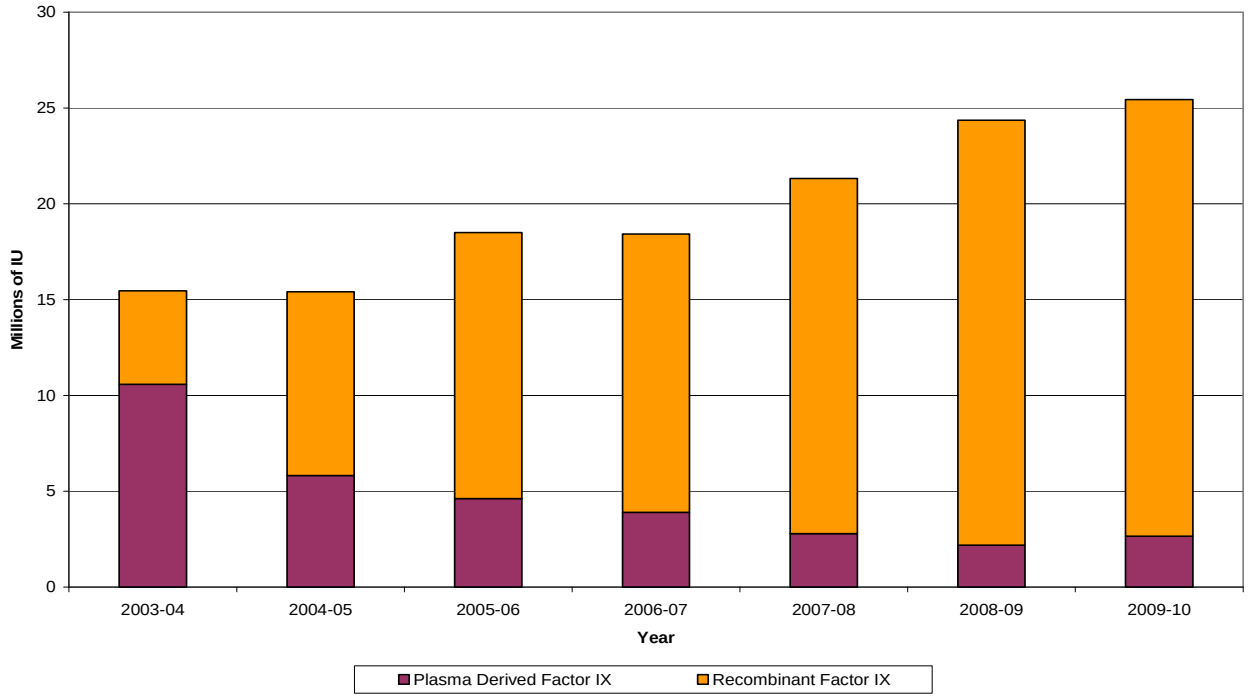
It will be important to monitor this slowing growth rate against the change in age distribution of severe patients depicted at Figure 15 and the model developed at figure 16.

Figure 18 Annual international units of Factor VIII issued nationally per 1000 population 2003-04 to 2009-10



Source: IDMS database of issues.

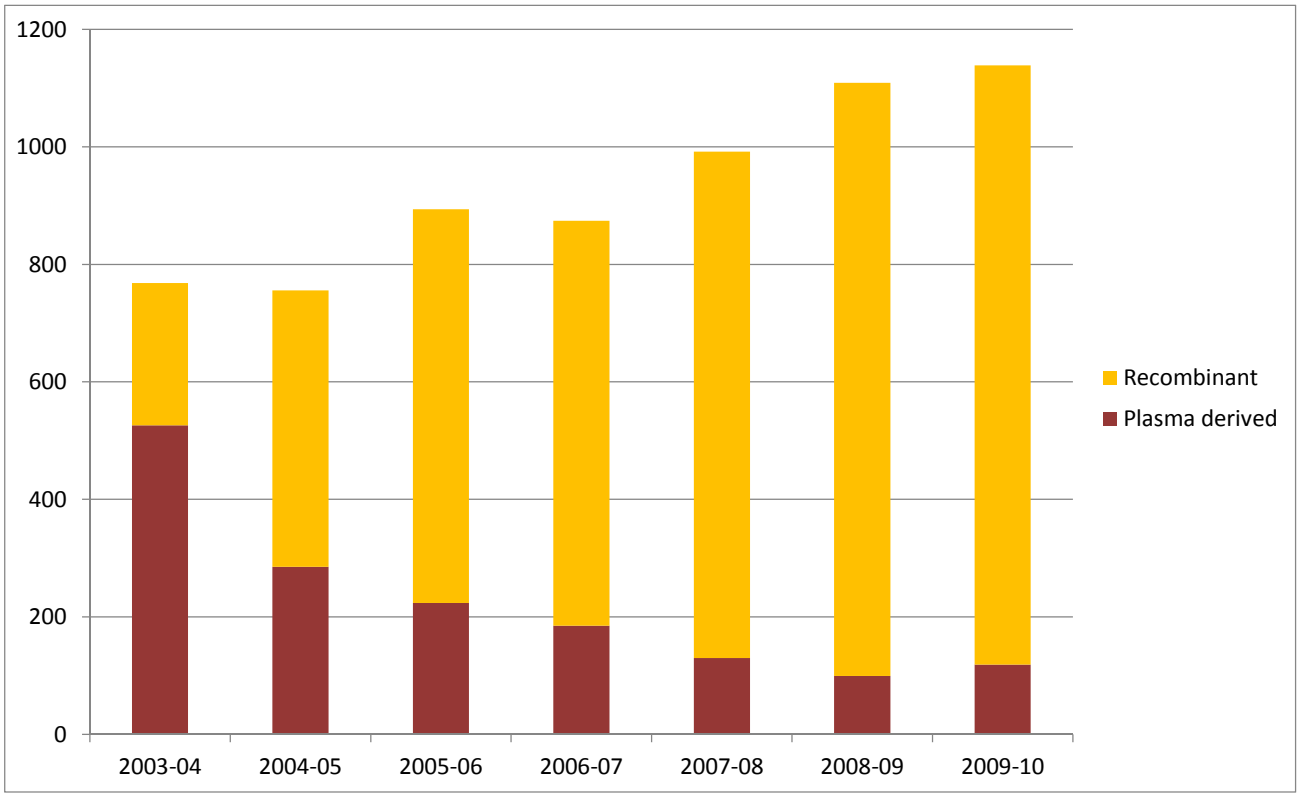
Figure 19 Annual international units of Factor IX issued nationally 2003-04 to 2009-10



Source: IDMS database of issues.

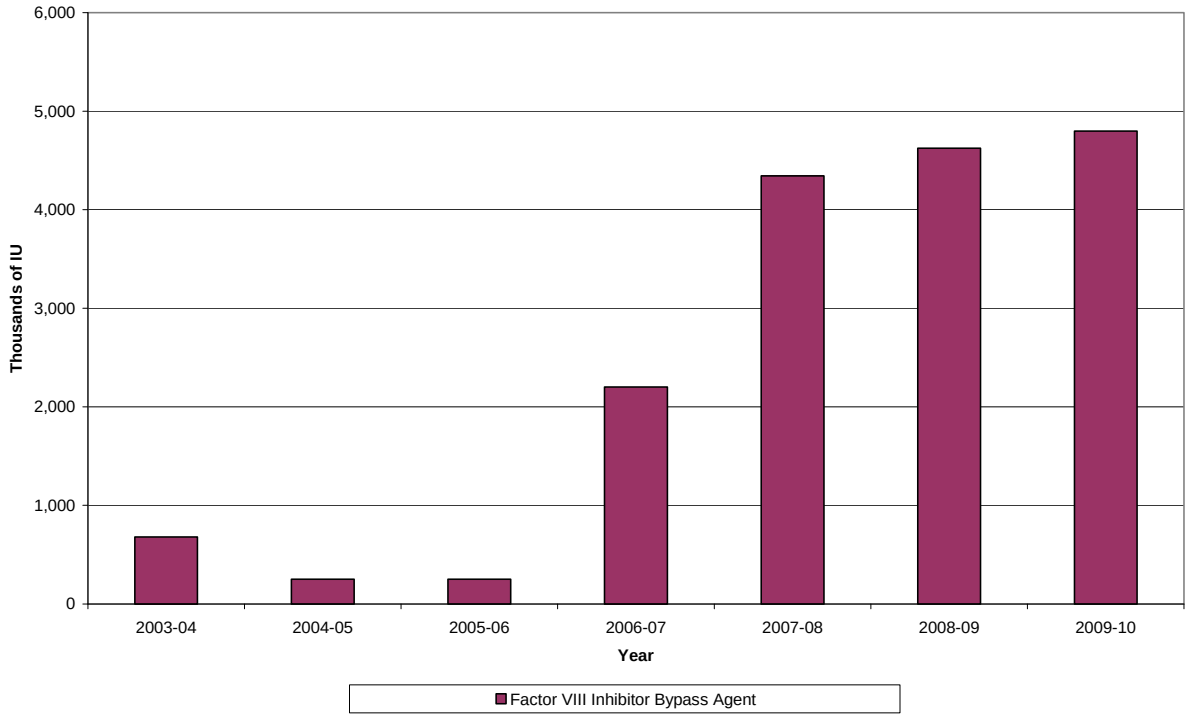
Similar to Figure 17, Figure 19 shows the strong growth of factor IX use (64.5 per cent or 8.6 per cent per annum) and the substitution of recombinant for plasma derived product.

Figure 20 Annual international units of Factor IX issued nationally per 1000 population 2003-04 to 2009-10



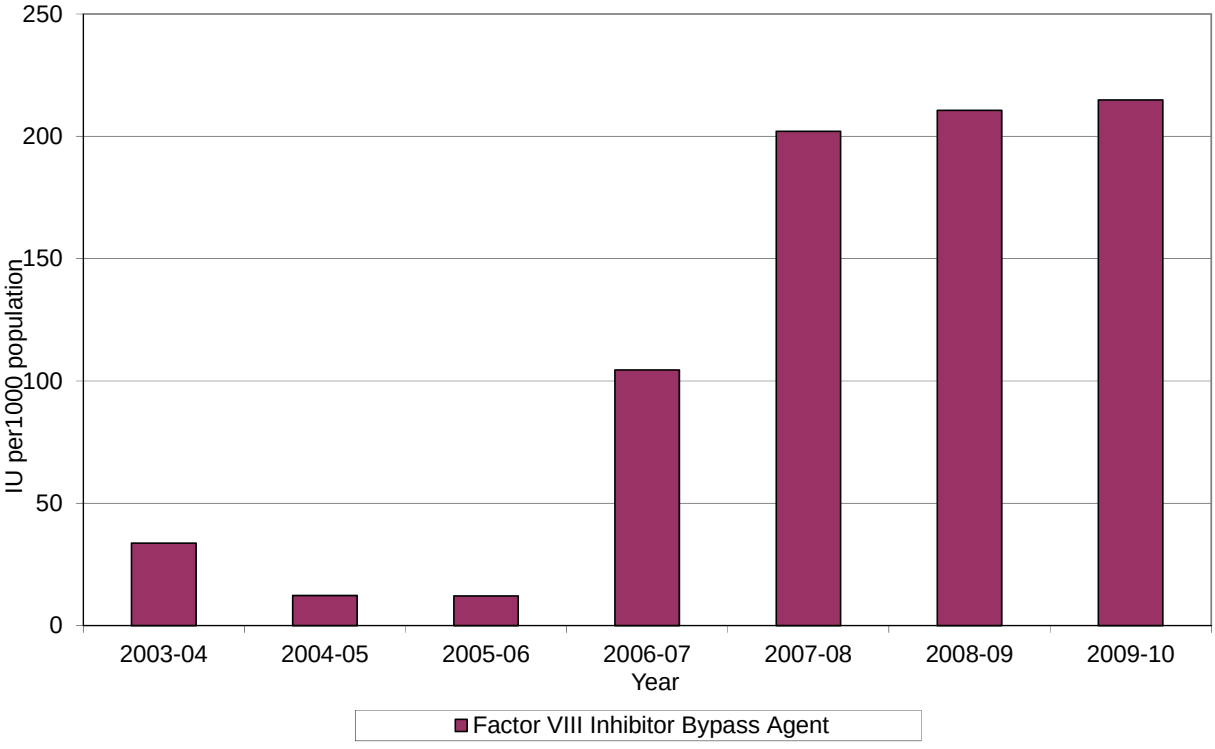
Source: IDMS database of issues.

Figure 21 Annual international units of Factor VIII inhibitor Bypassing Activity (FEIBA) issued nationally 2003-04 to 2009-10



Source: IDMS database of issues.

Figure 22 Annual international units of Factor VIII inhibitor Bypassing Activity (FEIBA) issued nationally per 1000 population 2003-04 to 2009-10



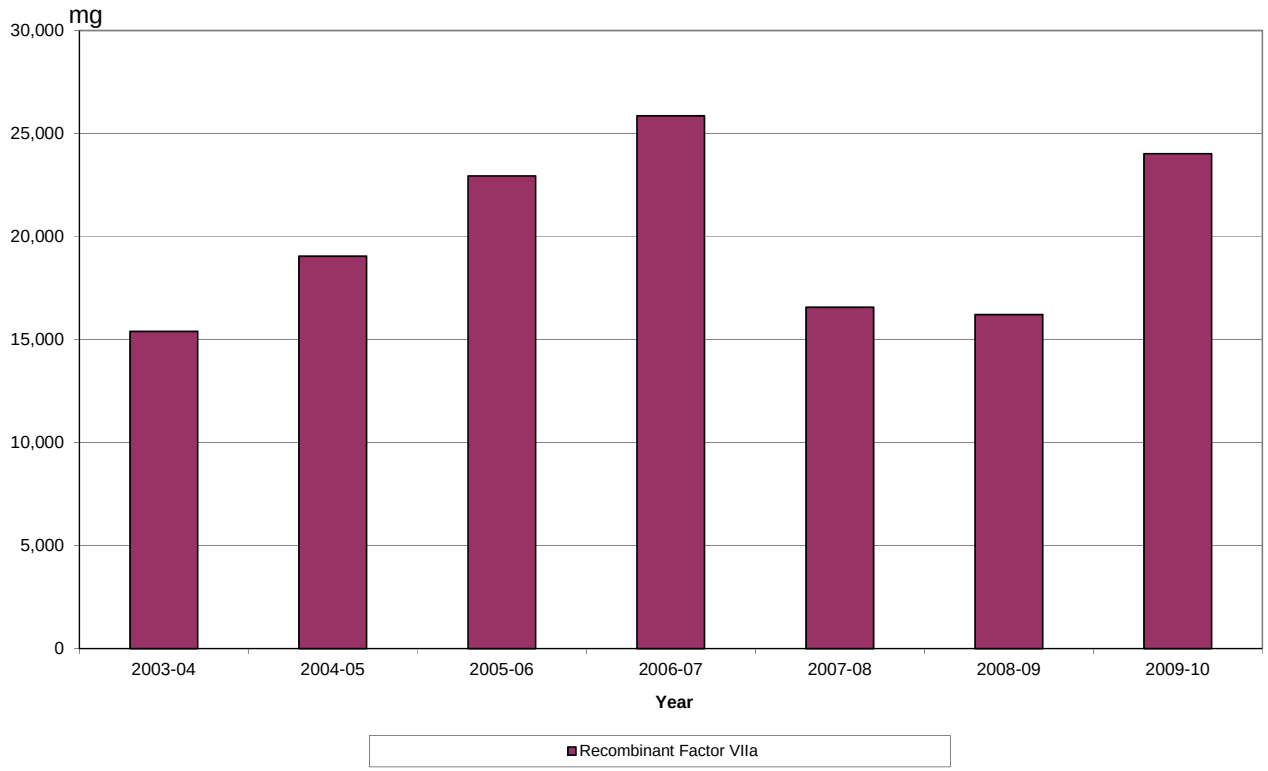
Since 2005-06 demand for FEIBA has increased significantly; in 2005-06 approximately 251,500 IUs were issued compared with almost 4.8 million IUs in 2009-10 (Figure 21). This represents an increase in growth of 775% in 2006-07 over 2005-06, and 97% in 2007-08 over 2006-07, although the growth in demand has slowed to 3.8% in 2009-10. Part of this growth relates to FEIBA becoming more widely available and government funded in 2006-07.

Predicting or interpreting changing demand trends is not possible with any accuracy, as the product is only used in a small number of patients each year. Use patterns will vary from year to year and will not only depend on the number of patients treated, but their severity of disease, the potency of inhibitors, whether secondary prophylaxis is practiced, the number and severity of spontaneous bleeds, and the amount of elective surgery undertaken in this patient group.

FEIBA and recombinant factor VIIa (rVIIa) (brand name NovoSeven) are both used to treat patients that have developed inhibitors.

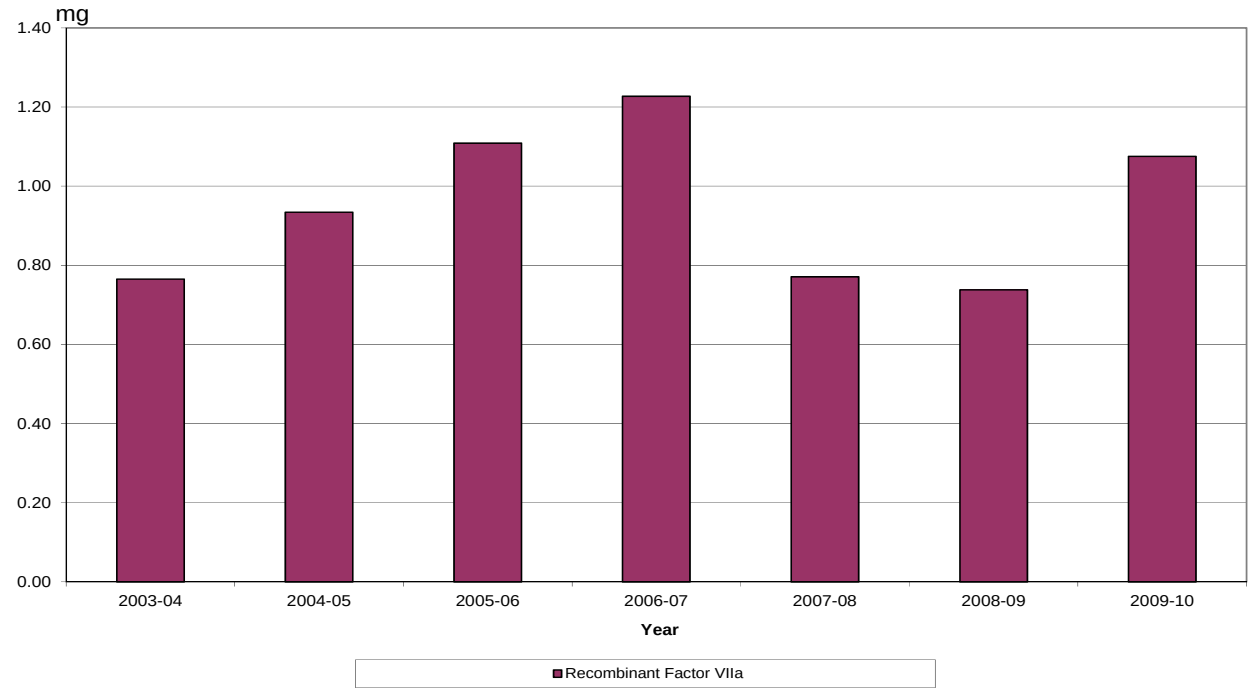
Drivers for clinical demand for FEIBA are similar to those for NovoSeven when issued under the National funding arrangements. The work of AHCDO in standardising protocols in haemophilia management will likely assist with driving a consistency of approach to the use of FEIBA and similar/related agents.

Figure 23 Annual international units of Recombinant Factor VIIa (NovoSeven) 2003-04 to 2009-10



Source: IDMS database of issues.

Figure 24 Annual international units of Recombinant Factor VIIa (NovoSeven) per 1000 population 2003-04 to 2009-10



Source: IDMS database of issues.

Demand for NovoSeven to treat patients with haemophilia under the National Blood Arrangements increased sharply from 2003-04 (15,400mg) to a high point in 2006-07 (25,900mg) but then declined to a more stable level in 2007-08 and 2008-09. However, high growth (47%) has been seen again from 2008-09 to 2009-10 (Figure 24). Demand for NovoSeven is very difficult to predict due to small patient numbers and patient specific requirements. At times very large doses can be needed by a single patient.

6.2. National Costs for products issued

Total expenditure on clotting factors for 2009/10 was $188.5 million. This was an increase of $14.9 million on 2008/09.

This represents about 24 per cent of the total blood and blood product budget for 2009/10. Expenditure by product is detailed in the NBA Annual Report available at www.nba.gov.au and is summarised below.

Table 16 National clotting factor costs 2008-09 and 2009-10 in $ million

	2008-09	**2009-10**
Factor VIII	104.3	109.3
Factor IX	26.1	26.7
Recombinant Factor VIIa	17.4	26.4
FEIBA	13.5	14.5
Other products	12.3	11.5
Total	173.6	188.5

7. Appendices

Appendix A Haemophilia Treatment Centres

The information at items 1 and 2 below is based on the original statement by the Australian Health Ministers Advisory Council which set out the intended objectives and benefits of the establishment of Haemophilia Treatment Centres (HTC's). The degree to which existing arrangements directly align to these original objectives varies.

1. The objectives of HTC's

Haemophilia Treatment Centres should be tasked with the provision of comprehensive care for people with haemophilia. Their roles can include:

- Compilation and distribution of guidelines for best practice directed toward optimal care of patients with disorders of haemostasis
- Providing protocols for the accurate diagnosis of patients with bleeding disorders
- Providing protocols for the regular review of infectious disease markers in patients and their families
- The allocation and distribution of therapeutic blood and recombinant products together with advice regarding the usage of blood and recombinant products, at a State and National level.
- The establishment of review programs to assess outcomes of therapies.
- Provision of regularly updated data to the national Haemophilia Register.
- Participation in basic and clinical research.

2. Operating concept

Haemophilia Centres coordinate and, where possible, integrate patient care, research and education to provide the optimal use of expertise and resources within hospitals and the community. One collaborative centre for each State and Territory may suffice but this must include adult and paediatric type centres.

The role of the Haemophilia Centres is to provide:

- a single point accountability for the care of patients with bleeding disorders with responsibility for the coordination, allocation and distribution of therapeutic resources for the treatment of patients, ie coagulation products derived either from blood donors or recombinant technologies
- a clinical service by experienced staff for persons with bleeding disorders and their families at short notice at any time of the day or night
- organisation of home therapy programs by the centre or in collaboration with other haemophilia treatment facilities.
- a counselling and advisory service for people with haemophilia and their families including genetic counselling and family planning.

- specialist medical expertise, principally haematology, surgery (the surgeons would have to be accredited to the Haemophilia Centre) rheumatology, infectious diseases and dental services.
- specialist allied health services to include physiotherapy, social work and podiatry.
- a laboratory service able to carry out all investigations required for the accurate diagnosis of haemophilia and other inherited disorders of the haemostasis and to have access, in association with other centres, to specialised testing facilities, for example gene typing.
- a system of record for all investigations, treatments, allocation of therapeutic products and adverse reactions.
- a capability to participate in research including clinical trials
- educational programs for medical staff, other personnel, patients and their families which promote care of patients with disorders of haemostasis.
- an outreach service to isolated patients and treating medical services. The outreach service may include:-
    - A haemophilia treatment facility located in a hospital that does not provide all the specialist services
    - Designated supervising medical practitioner

All isolated patients where care is managed in an outreach program should be registered at and be reviewed regularly by a Haemophilia Centre which would arrange delivery of and monitor the supply of therapeutic coagulation products.

Table 17 List of operational HTCs and their ID codes

#	Hospital	Haemophilia Treatment Centre	ID	State
1	The Canberra Hospital	Haemophilia Clinic	1TCH	ACT
2	Calvary Mater Newcastle	Haemophilia Treatment Centre	2CMN	NSW
3	Royal Prince Alfred Hospital	Haemophilia Treatment Centre	3RPAH	NSW
4	Sydney Children's Hospital	Centre for Children's Cancer and Blood Disorders	4SCH	NSW
5	The Children's Hospital at Westmead	Haemophilia Treatment Centre	5CHW	NSW
6	Royal Darwin Hospital	Haemophilia Treatment Centre	6RDH	NT
7	Royal Brisbane and Women's Hospital	Queensland Haemophilia Centre	7RBWH	QLD
8	Royal Children's Hospital	Queensland Haemophilia Centre Child and Adolescent Service	8RCH	QLD
9	Royal Adelaide Hospital	South Australia Haemophilia Treatment Centre	9RAH	SA
10	Women's and Children's Hospital	South Australia Haemophilia Treatment Centre	10WCH	SA
11	Royal Hobart Hospital	Tasmanian Haemophilia Treatment Centre	11RHH	TAS
12	The Alfred Hospital	Ronald Sawyers Haemophilia Centre	12TAH	VIC
13	Royal Children's Hospital	Henry Ekert Haemophilia Treatment Centre	13RCH	VIC
14	Fremantle Hospital	The Haemophilia Centre of WA	14FH	WA
15	Princess Margaret Hospital	The Haemophilia Centre of WA	15PMH	WA
16	Royal Perth Hospital	The Haemophilia Centre of WA	16RPH	WA
99	*Offshore Patient - Long Term*	*Offshore Patient - Long Term*	*99OPL*	
98	*Inactive Patients*	*Inactive Patients*	*98DPG*	

3. Data Quality of HTC data collections

Health professionals at HTCs are represented by:

- Australian Haemophilia Nurses Group (AHNG)
- Australia/New Zealand Haemophilia Social Workers' and Counsellors' Group (ANZHSWCG)
- Australia/New Zealand Physiotherapist Group (ANZHPG)

These representatives, along with the HFA have provided input into the initial design of the ABDR and continue to provide input from their respective areas of specialty.

The Data Managers at each HTC have elected Data Manager Co-Chairs. These Co-Chairs coordinate teleconferences, between all Data Managers, on a regular basis. The Data Manager Co-Chairs also have the functionality of raising issues, to the NBA, on behalf of their group. AHCDO has a major role in providing support to ABDR Data Managers through the funded model for Data Managers.

Data Quality initiatives implemented in 2009-10 include:

- Annual Data Manager Conference (In conjunction with HFA Conference)
- Data Manager Training Sessions
- Data Dictionary (a document that contains information about the data)project undertaken by ABDR Data Manager Co-Chairs)
- ABDR reporting suite (developed reports) for Haemophilia Treatment Centre Data Managers
- 'Advanced Search' functionality of the ABDR whereby Data Managers are able to extract information from the ABDR on an *ad hoc* basis to, for example, validate data entry and provide clinical information to HTC clinicians
- NBA financial support, through AHCDO funding, for HTC Data Managers
- The ABDR Newsletter is a functional tool that provides an update on issues such as changes to the ABDR and functionality enhancements. This update is a means of keeping all ABDR stakeholders informed. (Refer Appendix D for example of August 2010 ABDR Newsletter)

Data Manager Training Session – Melbourne 2010

Data Quality initiatives in 2009-10 have addressed:

- Clean up of migrated data
- Currency of data
- Unpopulated data fields
- National consistency of data quality
- Ongoing issues of information exchange when persons with haemophilia (PWH) move between states and haemophilia treatment centres
- Historical data (immediately prior to the deployment of the redeveloped ABDR in December 2008

Appendix B History and development of the ABDR

The ABDR was first established in 1988 using a 'Paradox' database at each Haemophilia Treatment Centre in Australia. The aims of the ABDR were to provide a clinical tool for improved management and national demographics of persons with haemophilia and other inherited bleeding disorders.

In 2000, a revised ABDR was established using 'Access' database platform at each Haemophilia Treatment Centre with a national collection of de-identified data every six months. Dedicated data base managers in individual centres improved data collection. Ongoing concerns regarding privacy prevented collection of national demographics such as age and gender.

To provide better sharing and access to the database it was decided in 2006 to move to an internet interface to central database. Genix Ventures was the successful tender with the National Blood Authority providing funding and project management.

The redeveloped ABDR was deployed in December 2008 at all HTCs.

1. Benefits of the redeveloped ABDR

The NBA redeveloped the ABDR and deployed the redeveloped ABDR in December 2008. It provides the following benefits:

- Single point of access for clinicians for treatment of patients
- Patient information relating to all clinical information associated with the treatment of haemophilia
- Information exchange between states and Haemophilia Treatment Centres
- National demographic information (age, gender etc) of persons with bleeding disorders
- National data on inhibitor incidence and outcomes of treatment
- Allied health (physiotherapy and social work) monitoring and outcomes
- Recording of personal usage of factor replacement for clinical monitoring
- Data for forward planning and funding of factor concentrates on a national basis
- High usage patterns

2. Current position of the development of the ABDR

Today the Australian Bleeding Disorders Registry is fully operational. The ABDR Steering Committee continues to oversee the project.

The National Blood Authority's role continues around provision of resources to maintain ABDR operations and to ensure timely and accurate reporting from the ABDR through provision of support to Data Managers. Data Managers, funded and supported by AHCDO, are located at HTCs across Australia.

Appendix C ABDR data collection (registration form)

ABDR
Australian Bleeding Disorders Registry

ATTENTION: ABDR DATA MANAGER

#bleeding disorder

Factor II deficiency (Prothrombin)
Factor V deficiency
Factor VI deficiency
Factor VII deficiency (Haemophilia A)
Factor IX deficiency (Haemophilia B)
Factor X deficiency
Factor XI deficiency
Factor XI deficiency
Factor XII deficiency
Symptomatic Carrier Factor VIII deficiency (Haemophilia A)
Symptomatic Carrier Factor IX deficiency (Haemophilia B)
Asymptomatic Carrier Factor VIII deficiency (Haemophilia A)
Asymptomatic Carrier Factor IX deficiency (Haemophilia B)
von Willebrand Disease Type 1
von Willebrand Disease Type 2 – Uncharacterised
von Willebrand Disease Type 2A
von Willebrand Disease Type 2B
von Willebrand Disease Type 2M
von Willebrand Disease Type 2N
von Willebrand Disease Type 3
von Willebrand Disease – Uncharacterised
Fibrinogen – Afibrinogenemia
Fibrinogen – Hypofibrinogenemia
Fibrinogen – Dysfibrinogenemia
Fibrinogen dysfunction – Uncharacterised
Platelet – Glanzmann's thrombasthenia
Platelet – Bernard-Soulier
Platelet – May Hegglin
Platelet – Macrothrombocytopenias
Platelet – Storage pool (dense granule) deficiency
Platelet – Primary secretion defect
Platelet – Uncharacterised
Acquired factor VIII inhibitor (Acquired Haemophilia A)
Acquired von Willebrand's Disease
Vascular disorders – Ehlers Danlos Syndrome
Vascular disorders – Uncharacterised
Other, please specify

+treatment regimen

On demand
Prophylaxis
Tolerisation
Secondary Prophylaxis

^Product Name (type)

Advate® (rFVIII)
Fresh Frozen Plasma (FFP)
BeneFIX® (rFIX)
Biostate® (pdFVIII)
Ceprotin® (Protein C)
Cryoprecipitate
DDAVP (Synthetic hormone)
Factor Eight Inhibitor Bypass Agent (FEIBA®) (Bypassing Agent)
Factor VII Concentrate® (pdFVII)
Factor XI bpl® (pdFXI)
Factor XI LFB Hemoleven® (pdFXI)
Fibrogammin P® (pdFXIII)
Fresh Frozen Plasma (FFP)
Haemocomplettan P 1g (pdFXIII)
Intravenous Immunoglobulin (IVIg)
MonoFIX® - VF (pdFIX)
NovoSeven® (rFVIIa)
NovoSeven RT® (rFVIIa)
Platelets
Prothrombinex™ - VF (pdPCC)
Recombinate® (rFVIII)
ReFacto® (rFVIII)

ABDR Patient Pamphlet

What is the ABDR? The Australian Bleeding Disorders Registry (**ABDR**) is a database that collects all clinical information related to the treatment of people with bleeding disorders, like an electronic medical file. This includes information about patient diagnosis, treatment details, hospital admissions and administrative information as well as details on ordering, supply and use of clotting factor products. Information is entered into the ABDR by staff at haemophilia treatment centres. The ABDR is managed by a service provider engaged by the National Blood Authority. The ABDR was first established in 1988 and has been upgraded many times with the latest significant upgrade in 2008.

Why do you need it? The ABDR provides your health care team and support staff with a record enabling them to monitor and manage your treatment over time to improve your quality of life. Depersonalised information available from the ABDR may be used by authorised organisations to understand and improve treatment for bleeding disorders. The ABDR also provides governments with information on total clotting factor product requirements to make sure there is enough available to meet the needs of all Australians with bleeding disorders.

What about privacy? Only the health care team and support staff involved in providing medical services to you have access to your personal information. Other authorised users only have access to limited, depersonalised and/or summary information where all identifying information is removed to protect your privacy.

Does information about me have to be included? A minimum amount of information about you is required to ensure the continuous supply of clotting factor product is available to meet your treatment needs.

Where can I get more information? Further information about the ABDR can be obtained from the Australian Haemophilia Centre Directors' Organisation (AHCDO) on (03) 9885 1777, email info@ahcdo.org.au or visit www.ahcdo.org.au

Endorsement from Haemophilia Foundation Australia

Haemophilia Foundation Australia supports the ABDR. It helps doctors and other treating health professionals to understand more about the care and treatment needs of people affected by bleeding disorders. The ABDR will assist and guide planning to ensure treatment product is available when it is needed. We are confident the steps in place will mean accurate, reliable and confidential data is available and that no patient details can be identified outside haemophilia centres.

www.haemophilia.org.au

Endorsement from Australian Haemophilia Centre Directors' Organisation

The ABDR is a valuable tool that provides an overview of those affected with haemophilia and other bleeding disorders in Australia. Data from the ABDR is the best information available for clinicians to advise governments making policy decisions regarding treatment needs and product availability.

National statistics available through the ABDR will give AHCDO an overview of practise and allow opportunities for improvement. This data can be pooled to compare Australian treatment standards with international benchmarks. The ABDR will continue to provide the ability to assess quality of life and other important clinical questions arising across Australia.

AHCDO's partnership on this initiative with the National Blood Authority, Haemophilia Foundation Australia and other specialist health professional groups is vital to the pursuit of excellence in clinical treatment practices.

www.ahcdo.org.au

Copies of this pamphlet can be obtained by contacting the ABDR Secretariat at abdr@nba.gov.au or 02 6211 8311.

When complete fax to your nearest Treatment Centre or Clinic – see www.ahcdo.org.au for details

Effective April 2009

Appendix D August 2010 ABDR Newsletter

ABDR
Australian Bleeding Disorders Registry

National Blood Authority Australia

Australian Bleeding Disorders Registry Newsletter
October 2010

ABDR User Survey

Thank you to all of you who have responded already to the ABDR User Survey. So far, over 40% of users have responded and the feedback received has already helped inform the deliberations of the Steering Committee.

If you have not yet responded, please take five to ten minutes of your time to respond. The link to the survey has been previously emailed to you. If you did not receive the email or need any other assistance, please contact the NBA for assistance.

A summary of the results from the survey will be contained in the November issue of the ABDR Newsletter.

2009-2010 ABDR Annual Report

The quantity and quality of the data in ABDR has continued to grow since its launch in December 2008 is now providing a meaningful source of information for extraction, analysis and reporting.

Barbara Herden in the NBA has been working on the production of the 2009-2010 ABDR Annual Report over the last few months.

The report covers the 2009-2010 financial year and contains a variety of patient de-identified data and analysis. The report is grouped into the following key areas:

- Data from a patient focus;
- Data from a supply focus including other NBA data;
- Analysis of the data, including where possible a comparison to historical data and benchmarking against comparable international data-sets; and
- ABDR governance

The report will be released online in late 2010 once it has been finalised and its release approved by the ABDR Steering Committee.

Key Contacts

- **Genix Ventures Support Desk**
  (Technical Issues)
  Phone – 13 000 BLOOD (13 000 25663), Option 1
  Email – support@csc.genixventures.com
- **National Blood Authority (NBA)**
  (Data. Governance & Project Management Issues)
  Phone – 13 000 BLOOD (13 000 25663), Option 9
  Email – abdr@nba.gov.au

Data Managers' Meeting

The Data Managers' met in Melbourne on 7 and 8 October 2010 along with representatives from AHCDO and the NBA to discuss a range of issues including the ABDR Annual Report, Advanced Search, data quality and the role of the DMs.

Thank you to Julia Ekert, Linda Mason and Megan Sarson for their wonderful organisational abilities and for their lively contributions to the deliberations of the group.

A number of key outcomes, relevant to all ABDR users were agreed at the meeting:

- Removal of all 'Other, Please Specify' diagnoses to be completed by 31 December 2010;
- All data back to 1 July 2008 to be fully populated and cleansed by 30 April 2011;
- A renewed emphasis to obtain (where possible) and record accurate patient weights in ABDR;
- The National Blood Authority in conjunction with the Co-Chairs of the Data Managers' to develop a draft data dictionary for consideration by the next meeting of the Data Managers'; and
- Agreement on the need to establish a handbook for Data Managers' that contains all of the information and guidance that a Data Manager could need to undertake their duties in ABDR.

New Password Reset Protocol

The Steering Committee has recently approved the introduction of a new password reset protocol to enhance the security of ABDR and to reduce the likelihood of anyone other than an approved user from resetting their password.

The process is being expanded to include a series of challenge and response questions whereby you will be asked to nominate in advance three questions and matching answers that relate to something that would not be readily known by anyone trying to gain unauthorised access to ABDR. Examples of such questions could include, the colour of your first car, the name of your first gold-fish etc.

Should you forget your password or lock out your account from 1 November 2010 onwards, you will be guided through the following password reset process by the Genix Ventures Support team:

1. Once you have called Genix Ventures, they will require you to answer each of the following correctly:
    a. Your full name
    b. Your HTC
    c. Your role (as per your login to ABDR)
    d. Your ABDR username
2. You will then be asked two of the previously supplied questions and you must respond with the correct answers to each.
3. You will be asked to re-confirm your email address and a temporary password will then be emailed to you.

I will be writing to each user in the coming week to securely seek their challenge and responses to provide to Genix Ventures by 31 October to enable the new arrangements to commence on 1 November 2010.

Excel Tips and Tricks

When extracting data from ABDR there is often a need (particularly when extracting using the Advanced Search Function) to manipulate data using Excel.

Whilst nothing can replace face to face training and support for such issues, there are a couple of freely available training sites on the web for Excel that may be of interest to users:

- www2.rmit.edu.au/departments/rp/excelhelp.php
- office.microsoft.com/en-au/excel-help/

Diagnosis – Other, Please Specify

One of the ways in which ABDR coped with the mass import of records from the old (ie pre December 2008) Access Database versions of the system was the creation of an 'Other, Please Specify' option in the 'Diagnosis' list of values.

Whilst such a solution was an ideal way to capture the varying ways in which diagnosis was recorded across HTCs, it is now causing issues when generating reports and statistics from ABDR and when using the Advanced Search module.

With the clean-up of such migrated records now well underway and nearing completion in some HTCs, it is an appropriate time to remove this option from the list of values for this field.

Effective immediately, all users should cease using the 'Other, Please Specify' option in the 'Diagnosis' list of values. Data Managers in each HTC will be editing all patient records where this value is used to select the diagnosis from the pre-existing list of approved conditions. This work will be completed by 31 December 2010 at which time Genix Ventures will remove the option from the list of values for the field.

Virtual HTCs

A second, virtual HTC, was recently established in ABDR to provide a suitable repository for inactive patients. This site is similar to the 'Overseas Patients – Long Term (99-OPL)' site that was created some months ago.

Data Managers in all HTCs are able to transition patients into both of these virtual HTCs. Their role is solely to provide a readily identifiable place to park patient records to assist in maintaining the integrity and usefulness of the data in ABDR.

Inactive Patients – Long Term (98-DPG)
This virtual HTC provides a space to 'park' the records of patients who are no longer receiving treatment (or product) from an Australian HTC.

Patients may only be transitioned to this HTC if they have not received treatment from the HTC at any time in the last three years.

If at any time a patient who has been transitioned into this virtual HTC receives treatment from an Australian HTC, they are to be immediately transitioned to that HTC.

All HTC Data Managers have access to this site and are able to transition patients to and from this HTC.

Overseas Patients – Long Term (99-OPL)
This virtual HTC provides a space to 'park' the records of patients who have moved overseas for a period of months or years (such as postings for diplomats or military officers) and during this time are not receiving treatment (or product) from any Australian HTC. This HTC is not to be used for patients that are receiving product overseas subsidised under the national blood arrangements.

The configuration of this site will not permit product orders, patient treatment cards, patient diagnosis or other interaction events to be raised for patients whilst they are located within the HTC.

The NBA act as Data Managers for the site with our role limited to facilitating transition between this site and other HTC sites when patients return (if they do).

ABDR Response Times

One of the most common issues raised relating to ABDR is the speed with which the system displays screens and reports. Part of this relates to the actual operations of the system itself, and there is little that can be done at present to increase the response times of the actual system.

Two other factors that affect the speed with which ABDR screens and reports are displayed are:

Your Browser
ABDR does not operate at its full speed when used on an old web browser (such as Internet Explorer 6).

If you are able, you should upgrade to a newer version of either Internet Explorer (version 7) or Mozilla Firefox (version 3.6). You should notice an immediate improvement.

Your Internet Connection
No matter how fast ABDR or your computer may be, if the internet connecting the two is slow, your ABDR session will be slow.

If you are experiencing an unusually slow ABDR session, you should run a test to identify if your internet connection speed may be the cause. There are a variety of free speed tests on the internet – one that I use is available at www.speedtest.net . Make a note of the download, upload and ping results and contact Genix Ventures to report the issue (and your results).

Use of ABDR Data

The ABDR Steering Committee at its meeting in December 2009 confirmed that HTCs were free to use data derived from ABDR within their own HTC for the purposes of managing the HTC and patients. Further, they confirmed that the data should be managed within the existing frameworks governing patient records in each hospital.

The Committee reaffirmed the requirement for HTCs seeking to use data from ABDR outside their HTC (for example in a conference presentation) to seek approval from the Steering Committee prior to the publication/release of the data. Nathan Kruger from the NBA is able to provide guidance on the process to seek this approval.

Maintaining ABDR Security

As an ABDR user, you will have been allocated and issued with a unique username and password which are used to logon to the ABDR. You must keep these logon details secure and not disclose them to any other person (other than to authorised ABDR support personnel for proper system or user administration purposes) at any time.

Supplier Contact Details

If you notice that any of the contact details for suppliers listed in ABDR have changed and we have not updated the system to reflect the change, please advise Nathan Kruger at the NBA so that we can correct the details for you.

Newsletter Back Issues and Topics

If you think you have missed out on one of our previous issues, or perhaps you are new to ABDR, feel free to contact the NBA to obtain a complete set of back issues of this newsletter.

If there is anything that you would like to see covered in the next issue of the newsletter, please let us know.

Peter O'Halloran
Chief Information Officer, National Blood Authority
13 October 2010

Phone: 13 000 BLOOD (13 000 25663), Option 9
Email: abdr@nba.gov.au

Appendix E

List of Figures

List of tables