Australian Bleeding Disorders Registry (ABDR) – An Update

Rowell, John[1]; Caris, Sharon[2]; Gunn, Stephanie[3]; Simon, Geoff[3]; Barnes Chris[1]

1. Australian Haemophilia Centre Directors' Organisation (AHCDO), Melbourne Vic *2. Haemophilia Foundation Australia (HFA), Melbourne Vic* *3. National Blood Authority (NBA), Canberra, ACT*

Background

The current ABDR is an internet based clinical registry including demographic, clinical and factor usage and complications data of those persons with inherited bleeding disorders registered. The project is managed by the ABDR Steering Committee (ABDRSC). Representatives are drawn from the major stake-holders in the project:-

AHCDO– a group of clinicians responsible for haemophilia care in Australia
HFA – Australia wide patient support group
NBA – federally funded agency responsible for procurement and supply of blood and recombinant products used in management of inherited bleeding disorders (IBD).

In 2008, following an extensive consultation with stakeholders and users, a tender was let to a commercial software vendor, with implementation of the product in 2009. There were significant issues in implementation including browser compatibility, speed, reliability of the application, report generation, data acquisition and data entry. In 2011 a decision was made to move to an industry standard platform rather than a proprietary database – in conjunction with further development with the NBA of internal IT support.

A project team oversees the development of software in a "like for like" format to take advantage of existing format and experience of data managers and other staff. The 4th generation ABDR is to be released in July 2012.

The 4th generation development has resulted in a more robust platform, correction of errors, improved logical data entry and additional screens involving physiotherapy, social work and adverse events of therapy.

Data collected can be reported on a National and State basis for review by individual Haemophilia Treatment Centres.

The Governance Framework for the ABDR is strictly compliant with Privacy and Freedom of Information Legislation. The ABDRSC has oversight for release of data for public reporting.

Data from the 2010/11 ABDR Report

Distribution of numbers registered in ABDR from June 2009 to July 2011

	Number in register at 30 Jun 2009	Number who Received product in 2008-09	Number in register at 30 Jun 2010	Number who Received product in 2009-10	Number in register at 30 Jun 2011	Number who Received product in 2010-11
HmA	1918	693	2015	833	2111	858
HmB	466	153	490	186	517	185
vWD	1714	101	1856	183	1966	153
Other Factor Deficiency	235	21	261	18	284	22
Platelet Disorder	161	1	170	4	191	8
Vascular	5	0	5	0	6	0
Other	138	2	143	0	146	2
Unknown	35	0	37	0	39	0
Total	**4672**	**971**	**4977**	**1224**	**5260**	**1228**

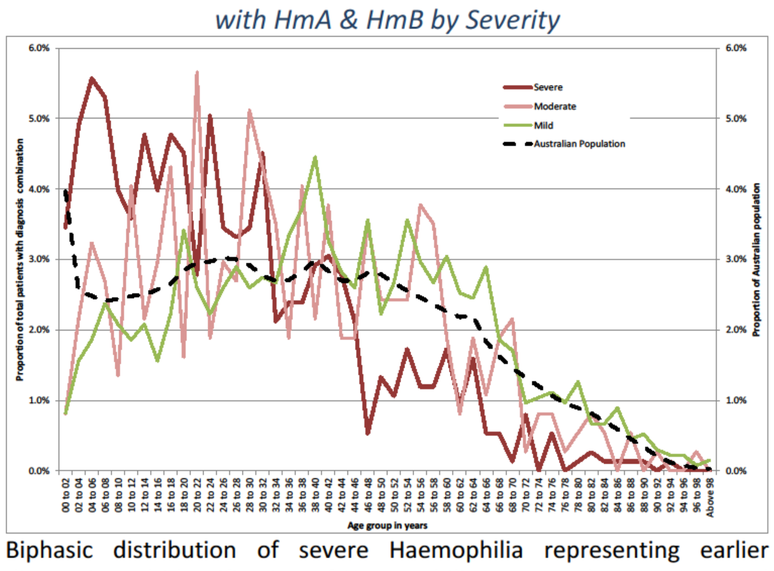
There has been a progressive increase in numbers of affected individuals in the last three years, representing 'real' increase in persons with IBD.

Distribution of numbers based on paediatric (<19 years) & adult (>19 years)

	Paediatric 0 – 19 years		Adult 20 years and over		Total	
	Number in register at 30 Jun 2011	Number who Received product in 2010-11	Number in register at 30 Jun 2011	Number who Received product in 2010-11	Number in register at 30 Jun 2011	Number who Received product in 2010-11
HmA	587	334	1524	513	2111	847
Severe	289	242	362	256	651	498
Moderate	71	46	190	83	261	129
Mild	219	44	856	171	1075	215
Not Applicable	1	0	29	2	30	2
Unknown	7	2	87	1	94	3
HmB	125	57	392	126	517	183
Severe	49	37	54	38	103	75
Moderate	23	16	88	38	111	54
Mild	50	4	226	49	276	53
Not Applicable	1	0	3	0	4	0
Unknown	2	0	21	1	23	1
vWD	446	29	1520	122	1966	151
Total	**1158**	**420**	**3436**	**761**	**4594**	**1181**

Approximately 55% of all paediatric patients receive therapy and 30% of adult persons receive therapy. The majority of therapy is provided to severe persons. Not applicable/Unknown represented continuing data entry challenges.

Distribution by age - people in the registry as at 30 June 2011 with HmA & HmB by Severity



Biphasic distribution of severe Haemophilia representing earlier diagnosis and identification of Haemophilia at an early age compared to the general population and possible effects of early mortality of severe Haemophilia due to bleeding or consequences of viral infections.

Number of patients with inhibitors

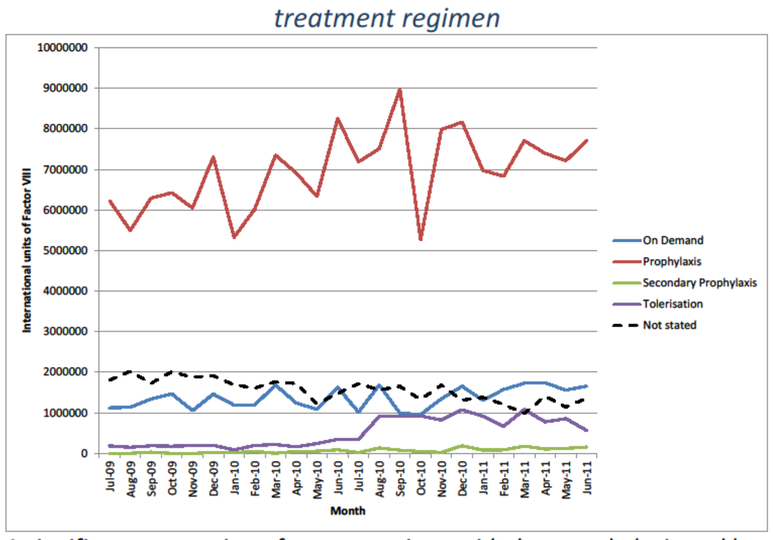
	Severe			Moderate			Mild			Total	
	Inhibitor present	proportion of total with inhibitors	Total Severe	Inhibitor present	proportion of total with inhibitors	Total Moderate	Inhibitor present	proportion of total with inhibitors	Total Mild	proportion of total with inhibitors	Total in Register at 2011
HmA	133	20.4%	651	15	5.7%	261	37	3.4%	1075	9.3%	1987
HmB	7	6.8%	103	0	0.0%	111	0	0.0%	276	1.4%	490
vWD	2	1.4%	138	0	0.0%	251	0	0.0%	1577	0.1%	1966

† For vWD severity is not always defined. Included in the mild vWD are those with severity 'mild', 'not applicable' and 'unknown'.
‡Inhibitors are usually only seen in type 3 (severe) VWD - so if the number of type 3(31) were to be used then becomes similar to UK proportion

Proportion of those with inhibitors is similar to reported in 'UKHCDO Annual Report 2010/11'. Further analysis is required to identify risk factors including genetic mutation, those with active inhibitors and response to tolerisation and other therapies.

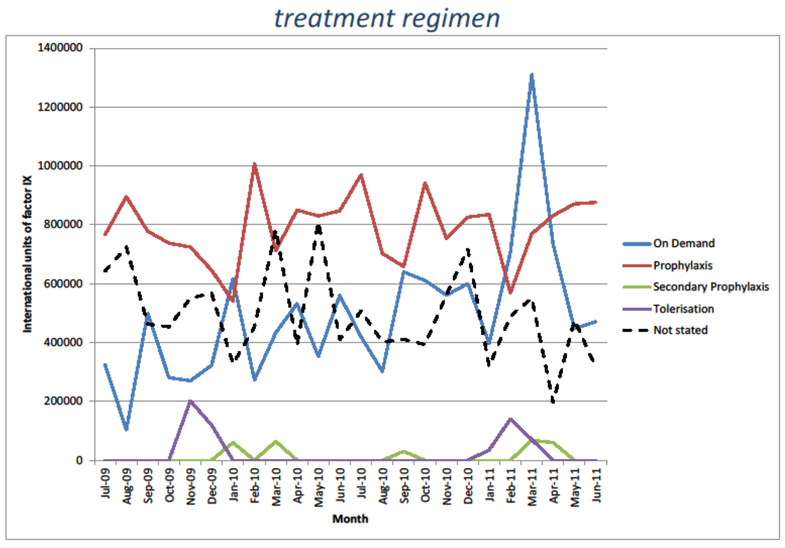
There is a significant group of those with moderate and mild Haemophilia A with inhibitors.

International Units of FVIII received by HmA patients by treatment regimen

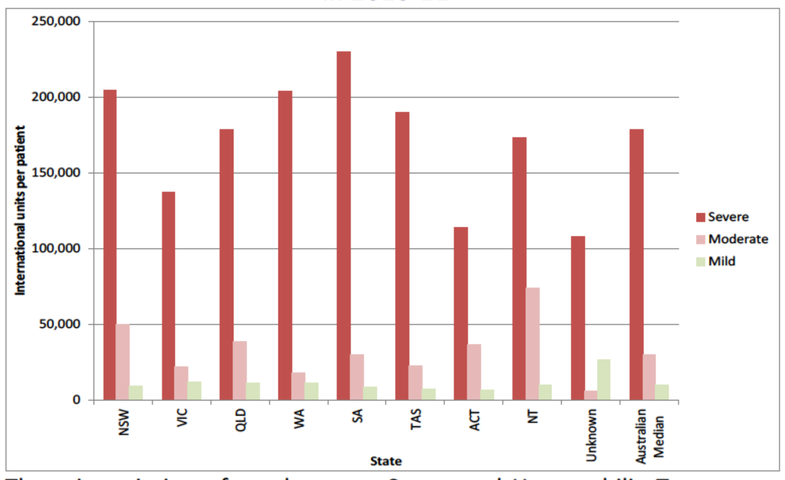


A significant proportion of treatment is provided as prophylaxis and has gradually increased over the last two years. Reports of tolerisation usage appears to have increased since 2010 but this may be related to increased reporting.

International Units of FIX received by HmB patients by treatment regimen

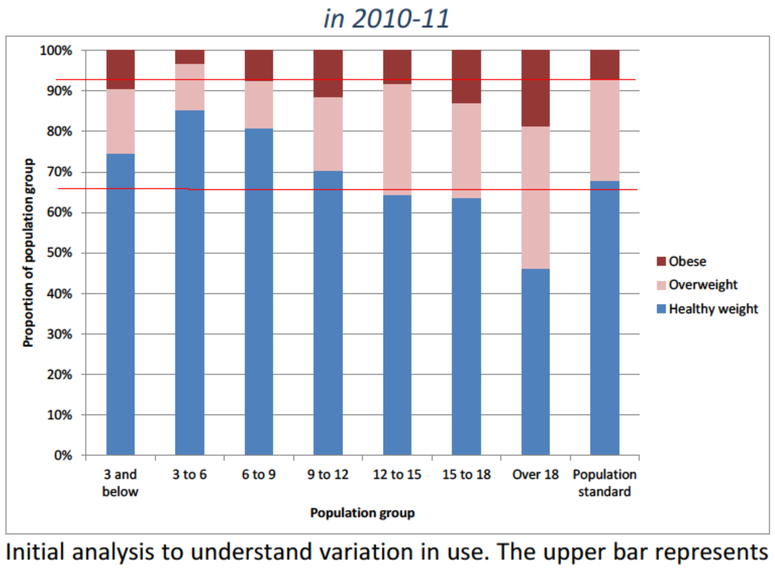


Median Factor VIII per HmA patient by jurisdiction and severity in 2010-11



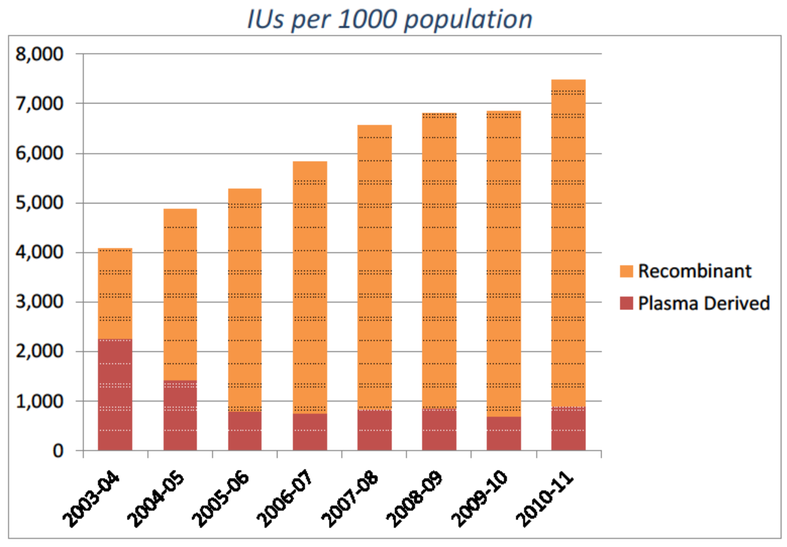
There is variation of use between States and Haemophilia Treatment Centres. Further analysis is required to identify those on tolerisation and analyse by age and severity. The unknown group represents those persons not seen at HTC – temporarily overseas.

Weight and height data for ABDR patients receiving treatment in 2010-11

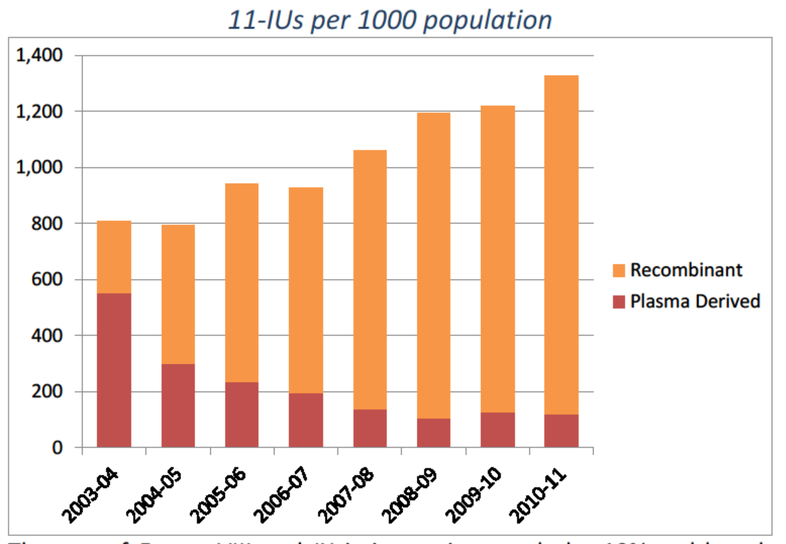


Initial analysis to understand variation in use. The upper bar represents general population levels of obesity and lower bar that of overweight. Further data analysis is required to clarify the effect of excessive weight on usage and outcomes.

Annual use of Factor VIII issued nationally 2003-04 to 2010-11– IUs per 1000 population



Annual use units of Factor IX issued nationally 2003-04 to 2010-11-IUs per 1000 population



The use of Factor VIII and IX is increasing yearly by 10% - although there was a pause in 2009/10. This will translate into increased costs and highlights urgent need to understand and manage demand while maintaining outcomes.

Future

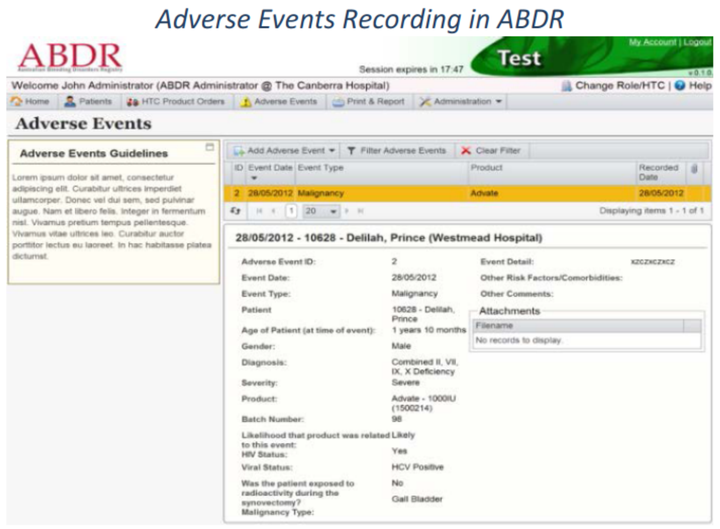
The ABDR has now been re-engineered to improve performance and adapted to meet changing needs of stakeholders

The ABDR has been enlarged to include physiotherapy assessment and social work assessment to enable and record the multidisciplinary approach to Haemophilia and other IBDs.

In development is a patient interface to allow direct usage data to be entered into the ABDR and include other parameters of response to treatment and effect on lifestyle or work.

The redevelopment will enable benchmarking of Haemophilia Centres and greater capacity to investigate the reasons for variation of replacement therapy and eventually the impact on those with Haemophilia. This investigative process will be further enhanced with the implementation of the ABDR Adverse Events recording. Clinical research will also be funded to further enhance stakeholder outcomes.

Adverse Events Recording in ABDR



Acknowledgements

The assistance of the following organisations:-

- *Australian Haemophilia Centre Directors' Organisation (AHCDO)*
- *Australian Haemophilia Nurses' Group (AHNG)*
- *Australia/New Zealand Haemophilia Social Workers' and Counsellors' Group (ANZHSWCG)*
- *Australian and New Zealand Physiotherapy Haemophilia Group (ANZPHG)*
- *Barbara Herden, National Blood Authority[3], for assistance for poster preparation*

DOI: 10.3252/pso.eu.WFH2012.2012