Understanding of the significance and health implications of asplenia in a cohort of patients with haemoglobinopathy: possible benefits of a spleen registry

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ABSTRACT

Objectives: Asplenia and hyposplenism carry a significant risk of ongoing morbidity and mortality which can be reduced by education, vaccination and antibiotic use. We aimed to assess education and other methods of prevention in a cohort of patients with haemoglobinopathy in a tertiary referral centre, which also had access to a post-splenectomy registry created to reduce post-splenectomy infection risk.

Methods: A standardized questionnaire was used on patients who attended the service for regular therapy. Patients were also asked about standard post-splenectomy preventive therapies including antibiotics and vaccinations.

Results: There were 49 patients who had either had a splenectomy or knew their spleen to be non-functional. Of these, nearly half knew themselves to be on the Victorian Spleen Registry (51.0%). The median knowledge score was 12 (range 4–17) out of a possible 18. Most significantly the benefits of the registry were not seen in terms of knowledge but in delivery of recommended vaccines and the use of a medical alert card.

Conclusion: This study examined knowledge and attitudes about splenectomy in a cohort of haemoglobinopathy patients in an Australian tertiary referral centre. The majority had good or fair knowledge with a strong association of some elements of post-splenectomy care with being placed on a spleen registry and having received targeted education. Implementation of systematic approaches by medical staff is likely to be the main benefit of a clinical registry approach in this setting.

Background

Asplenia and hyposplenism carry a significant risk of ongoing morbidity and mortality with an estimated annual rate of overwhelming post-splenectomy infection (OPSI) in the order of 1 in 500 [1–4]. This condition has a mortality rate of approximately 50% [2]. In addition, there is some suggestion that other infections may occur more frequently in this population compared with the general population [5–7]. The risks of missing splenic function may be mitigated by vaccination [8], prolonged antibiotic prophylaxis, the best evidence for which is in sickle-cell disease (SCD) [9], and education [1,7,8]. This three-pronged approach is recommended in guidelines for the care of those with absent splenic function [7,10].

Patient education is a simple and effective method of improving outcomes in many medical conditions, including the haemoglobinopathies, however there is little research in this area [11,12]. Patient knowledge about infection has previously been assessed in caretakers of children with SCD [12] as well as splenectomised populations. A study by Hegarty et al. in this latter group used a survey that included questions on vaccination and antibiotic prophylaxis [13]. The results demonstrated that involvement of a general practitioner (GP) in patient instruction was helpful in ensuring patients had an informed, active knowledge of their condition. Unfortunately, the statistical significance of this difference was not measured, and may have been inconclusive regardless given the small cohort size. The questionnaire has since been used in other papers to measure health literacy and understanding of asplenic patients [8,14].

In the State of Victoria, Australia, a spleen registry was formed in 2003 [15,16]. The Victorian Spleen Registry (VSR) records the names of asplenic and hyposplenic patients, and provides reminders about vaccination and antibiotic use as well as educational material [15,16]. This includes an educational kit provided on registration and an annual newsletter [17], which act as an adjunct to information provided by patients’ healthcare workers. As part of a wider study of infectious risks in a cohort of haemoglobinopathy patients at Monash Health, the state reference centre for adult thalassemia, we aimed to assess educational understanding and factors that may have played a role in it including membership of the spleen registry.

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Method

Questionnaire

The aim of the questionnaire was to determine the knowledge and attitudes of patients regarding infection prevention. In addition to some basic demographic information, the wider study covered the areas of vaccination uptake, chronic infections, animal contact, management plans and travel. An additional set of questions was included for patients who had undergone a splenectomy or knew themselves to have a non-functional spleen. These were taken from the study by Hegarty et al. with some additional questions, which included exposure to intravenous antibiotics and adherence to vaccination recommendations of the spleen registry.

Participant selection

The questionnaire was conducted in patients with a diagnosis of a haemoglobinopathy requiring treatment. They were identified by the haematology nurses and doctors working in our haematology department. Only adult patients were approached regarding participation in the study.

Recruitment

Patients attended the transfusion centre for therapy at regular. During these visits, patients were invited to participate in the study. If they consented to participate, the questionnaire was then conducted as a verbal interview, with patients given the opportunity to skip any questions they did not wish to answer.

Statistical analysis

Data was analysed as descriptive statistics using the help of the frequency and cross-tabulation functions. The chi-squared ($\chi^2$) test and Fisher’s exact test were used to assess differences between categorical variables. Odds ratios and 95% confidence intervals (CI) were also calculated. These parameters were compared between different groups using the Student’s t-test for independent samples.

Data points were considered to be outliers if they were more than three standard deviations away from the mean; these were excluded from all statistical calculations. All $p$-values calculated were two-tailed and, for each analysis, $p \leq .05$ was considered significant.

Results

There were 49 (57.0%) patients who had either had a splenectomy or knew their spleen to be non-functional (Table 1). Of these, nearly half knew themselves to be on the VSR (51.0%), with 13 (26.5%) saying they were not and 11 (22.4%) not knowing if they were. The responses are summarized in Table 2.

The median score was 12 (range 4–17) out of a possible 18. The distribution of scores is found in Figure 1 and summarized in Table 3. Table 4 shows their relation to the primary educating physician with respect to post-splenectomy care and Table 5 their relation to ongoing GP involvement. In terms of knowledge level there was no significant difference between those on the registry and those not on the registry with an odds ratio of 1.06 (95% CI 0.25–4.50)

Almost half of the patients claimed to have received the meningococcal vaccine within the last five years (49.0%), while 22.4% said they had not. However, nearly three quarters of patients did not know whether they had ever received the Haemophilus influenzae type B vaccine (73.5%). While this may have been affected by recall bias, responses were checked against the medical records where available.

Responses to the questions about intravenous antibiotics and adherence to vaccination recommendations of the spleen registry.

| Question                                                                 | No | Yes | Don’t know |
|-------------------------------------------------------------------------|----|-----|------------|
| 1. Are you aware of any effects asplenia has on your general health? Do you think it increases your chances of certain infections? | 2  | 15  | 32         |
| 2. Is there any way of preventing this? Do you know if you had any vaccinations? | 9  | 19  | 21         |
| 3. Is there anything else you can do to prevent infection? Are you taking antibiotics? | 3  | 9   | 37         |
| 4. How long will you take antibiotics? Will you take them for life? | 10 | 2   | 37         |
| 5. What if you are travelling far abroad? Is there a greater risk of malaria? | 25 | 22  | 2          |
| 6. What would you do if you got sick, e.g. catching a cold or flu? Would you take a full dose of antibiotics? | 17 | 17  | 15         |
| 7. Do you know the name of your antibiotics? | 6  | NA  | 43         |
| 8. Do you have up-to-date full dose antibiotics at home? | 9  | NA  | 40         |
| 9. What would you do if you got a scratch or a small dog bite? Would you visit your family doctor? | 18 | 7   | 24         |

Note: 0 = wrong/does not know; 1 = moderate passive knowledge; 2 = good active knowledge.
antibiotics and emergency antibiotics were similar, with 6 (12.2%) unsure in each case (Table 6).

Most patients did not have a medical alert or wallet card indicating that they had asplenia (71.4%). However, those that knew themselves to be on the VSR were significantly more likely to have one ($p = .007$) (Table 7).

The level of knowledge exhibited by patients varied between questions. The first question about the effects of asplenia was well answered, with only 2 patients out of 49 (4.1%) answering incorrectly; this suggests that patients are retaining information received about the consequences of splenectomy. The other questions that were well answered pertained to the use of antibiotic prophylaxis, indicating that this recommendation of the VSR was being implemented successfully. Most patients knew about the need to take lifelong antibiotics (39/49), knew the name of their antibiotic (43/49), and had a supply of it at home (40/49). Overall there was no difference between those on the registry and those not on the registry with regards to individual questions.

Most significantly the benefits of the registry were seen not in terms of knowledge but rather in delivery of recommended vaccines and the use of a medical alert card (Table 7), though because the numbers were low the benefit did not reach statistical significance for all parameters.

### Table 3. Quality of patients’ knowledge.

| Category of knowledge   | n  | %   |
|-------------------------|----|-----|
| Good knowledge (0–5)    | 15 | 30.6|
| Fair knowledge (6–13)   | 31 | 63.3|
| Poor knowledge (14–18)  | 3  | 6.1 |

### Table 4. Cumulative scores in relation to primary information source.

|                  | n | Good knowledge (%) | Fair knowledge (%) | Poor knowledge (%) |
|------------------|---|--------------------|--------------------|--------------------|
| Haematologist    | 28| 28.6               | 64.3               | 7.1                |
| Surgeon          | 6 | 50.0               | 50.0               | 0.0                |
| GP               | 1 | 0.0                | 100                | 0.0                |
| None             | 14| 28.6               | 64.3               | 7.1                |

### Table 5. Cumulative scores in relation to ongoing GP involvement.

|                  | n | Good knowledge (%) | Fair knowledge (%) | Poor knowledge (%) |
|------------------|---|--------------------|--------------------|--------------------|
| GP involved      | 37| 32.4               | 64.9               | 2.7                |
| GP not involved  | 12| 25.0               | 58.3               | 16.7               |

### Table 6. Responses to additional asplenia questions.

|                                | n | %   |
|--------------------------------|---|-----|
| Have you received the meningococcal vaccine in the last 5 years? | 24 | 49.0 |
| I don’t know                  | 11| 22.4|
| Have you ever received the Hib (Haemophilus) vaccine? | 5 | 10.2 |
| Yes                           | 5 | 10.2 |
| No                            | 8 | 16.3 |
| I don’t know                  | 14| 28.6|
| Have you ever received continuous IV antibiotics? | 20 | 40.8 |
| No                            | 23| 46.9|
| I don’t know                  | 6 | 12.2|
| Have you ever received emergency antibiotics? | 18 | 36.7 |
| Yes                           | 18| 36.7|
| No                            | 25| 51.0|
| I don’t know                  | 6 | 12.2|
| Do you have a medical alert medallion or wallet card that indicates your spleen is not functional or is absent? | 14 | 28.6 |
| Yes                           | 14| 28.6|
| No                            | 35| 71.4|
| Total                         | 49| 100 |

Figure 1. Distribution of cumulative scores for the Hegarty et al. questions.
**Table 7.** Effect of VSR status on uptake of VSR recommendations.

|                        | Meningococcal vaccine received | Meningococcal vaccine not received | Odds ratio (95% CI) | p-Value |
|------------------------|--------------------------------|-----------------------------------|---------------------|---------|
| On VSR Yes             | 15                             | 3                                 | 5.00 (0.93–26.79)   | .062    |
| On VSR No              | 6                              | 6                                 |                     |         |
|                        | Hib vaccine received            | Hib vaccine not received           |                     |         |
| On VSR Yes             | 4                              | 2                                 | 8.00 (0.50–127.90)  | .175    |
| On VSR No              | 1                              | 4                                 |                     |         |
|                        | Continuous IV antibiotics received | Continuous IV antibiotics not received |                     |         |
| On VSR Yes             | 13                             | 8                                 | 2.28 (0.54–9.67)    | .224    |
| On VSR No              | 8                              | 7                                 | 2.10 (0.47–9.30)    | .270    |
|                        | Emergency antibiotics received  | Emergency antibiotics not received |                     |         |
| On VSR Yes             | 12                             | 10                                |                     |         |
| On VSR No              | 7                              | 4                                 | 12.00 (0.95–115.66) | .007    |
|                        | Medical alert                   | No medical alert                  |                     |         |
| On VSR Yes             | 13                             | 12                                |                     |         |
| On VSR No              | 1                              | 12                                |                     |         |

**Discussion and conclusions**

This study examined knowledge and attitudes about splenectomy in a cohort of haemoglobinopathy patients in an Australian tertiary referral centre. It was undertaken prospectively in a well described cohort using a standard tool that has been used in other studies. The total number of patients was modest but likely to be typical of the patients sampled. The majority had good or fair knowledge with a strong association of some elements of post-splenectomy care with being placed on a spleen registry and having received targeted education.

The study did show the benefit of the use of a spleen registry and a targeted education programme even in patients chronically associated with health care. Surprisingly however the benefit came more from implementation of recommendations than knowledge of their condition, though the level of knowledge in the whole cohort was high when compared to overseas studies. This possibly demonstrates a cohort effect, and the benefits could be better quantified by using a number of cohorts in different geographical settings. In addition, this is a population with strong links to medical care because of their condition and may not be typical of all asplenic patients in many ways. However, it is noteworthy that at least 40% had received intravenous antibiotics at some stage. While this study did not look at a direct link between knowledge and severe bacterial infections given the low rate of OPSI, previous studies have shown that being in a registry is protective [14].

In the original study of Hegarty et al. 85% had good or fair knowledge of the implications of their condition [13], while in the El Alfy study 75% had good or poor knowledge [8]. Our figures are comparable to that despite no registry framework for education, however the delivery and adherence to guidelines of care in the registry are greater than reported elsewhere. This suggests the main benefit of the registry may be through medical rather than patient behaviour, and implementation of systematic approaches by medical staff is likely to be the main benefit of a clinical registry approach in this setting.

**Disclosure statement**

No potential conflict of interest was reported by the authors.

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