

Protein S Deficiency And Thrombophilia

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Title: Protein S Deficiency Research Survey (PSD1)

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Summary

Our survey asked people with Protein S Deficiency to give basic details of their health, lifestyle and experiences dealing with this rare blood disorder which is a recognised risk factor for venous thrombosis. The response indicates a strong interest from people affected by Protein S Deficiency to participate in further research and an opportunity for medical professionals to conduct follow up studies using a list of email contacts managed by www.protein.org.uk.

Introduction

Protein S Deficiency is a rare condition that can lead to deep vein thrombosis, pulmonary embolism or stroke and is often treated with long-term anti-coagulant therapy. On our web site at www.protein.org.uk we help people to find out about topics such as what proteins do, the different types of Protein S Deficiency (PSD), thrombophilia, anti-coagulants, blood tests and where to get Medic Alert bracelets. We have an online forum which provides an excellent opportunity for people with Protein S Deficiency to come together and discuss a wide range of issues.

The members of our online community frequently express an interest in finding out more about research and they also want to see if they can help in some way. Our survey demonstrates to the medical profession our willingness to help and will hopefully encourage an increase in information exchange between researchers and those affected by PSD. We have also established a mailing list so that it will be easier for researchers to contact the people who want to learn more about their research activities.

Methodology

We set out to collect the results of our survey using an online questionnaire hosted on our web server. We customised a cgi script so that it would take the responses and enter them into a delimited text file that could be easily imported into a spreadsheet making the processing of responses more straightforward. The last field of the questionnaire was used to record technical information including the remote host address, date, time and survey reference in case we encountered problems with the operation of the script.



Protein S Deficiency And Thrombophilia



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To comply with the Data Protection Act we formed an unincorporated members club which allows for the collection of personal information relevant to club activities. This is based on a Yahoo! Group which lets people opt-in to a mailing list and then receive instructions on how to access the online questionnaire. We promoted the opportunity to join this mailing list on our web site which has approximately 6,000 visits per month and nearly 400 forum members.

Members of the research mailing list were invited to take part in the online survey. 92 responses were collected between 10th September 2001 and 27th February 2002. Four responses were duplicated entries and one entry was considered incomplete leaving a total of 87 valid responses. 95% (84/87) reported a diagnosis of Protein S Deficiency (PSD).

Survey Results

The frequency of results is shown inside round brackets and each question is shown with the original text in full as follows:

Data Disclosure: Your participation in this research is voluntary. Disclosure of a medical condition could affect existing or future entitlement to health insurance or employment. If you choose to submit any information it is on the understanding of the following:

- You are a member of our Yahoo! Group (an unincorporated members club)
- You do not object to the data being held for this purpose
- You are aware that IP address and server log files are recorded and may be revealed under requirement of law. This is no different from using other web sites although it may allow you to be identified.
- You grant usage and indemnify the webmaster and third parties from any form of legal action connected with the collection, storage, retrieval, processing, publication and presentation of data submitted by you, either about yourself or anyone else.

People who disagree with any of the above points should not participate.

[01] Name

(87) Anonymous

These details were requested in case it became necessary to solve technical issues and we were asked to locate a specific entry.



Protein S Deficiency And Thrombophilia



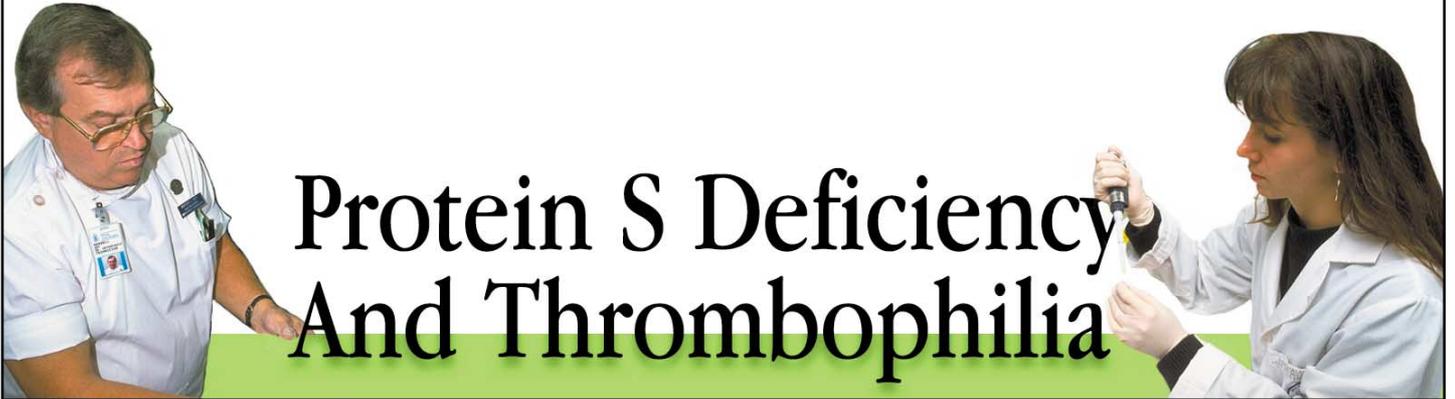
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[02] Age

- (4) Ages 0-9
- (3) Ages 10-19
- (12) Ages 20-29
- (28) Ages 30-39
- (16) Ages 40-49
- (13) Ages 50-59
- (5) Ages 60-69
- (3) Ages 70-79
- (1) Ages 80-89
- (2) No Response

[03] Relationship

- (2) Other Female
- (0) Grandmother
- (5) Mother
- (2) Sister
- (2) Daughter
- (46) Self-Female
- (10) Self-Male
- (4) Son
- (10) Brother
- (4) Father
- (0) Grandfather
- (2) Other Male



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[04] Does this person have Protein S Deficiency? We want to understand when people find out their PSD Status. If you were tested before having any health problem (e.g. alerted by a family member) please select 'Pre-tested'. If you learnt of PSD during routine screening (e.g. pregnancy or a company medical) select 'Screened'. If you learnt of PSD after an actual or suspected thrombosis select 'Diagnosed'. If you know what sort of PSD please also choose from 'Hereditary' or 'Acquired'.

- (32) Diagnosed - PSD
- (24) Diagnosed - Hereditary PSD
- (13) Pre-Tested - Hereditary PSD
- (5) Screened - Hereditary PSD
- (4) Diagnosed - Acquired PSD
- (3) Pre-Tested - PSD
- (3) Unknown
- (2) Screened - PSD
- (1) No Test Results

[05] Does this person have a history of Thrombosis? We want to understand if you have had more than one type of thrombosis. Use the abbreviations if possible (e.g. DVT, PE) but don't go into any other details.

- (41) DVT = Deep Vein Thrombosis; (21 DVT, 18 LDVT, 7 RDVT)
- (27) PE = Pulmonary Embolism
- (19) PB = Phlebitis
- (8) S = Stroke
- (7) PREG = Pregnancy - include this if it led to thrombosis
- (1) HA = Heart Attack
- (7) Enter the full name of the condition for any others not shown: clotting after surgery, ischemic colitis, underactive thyroid, acute vasculitis, retinal artery occlusion/blindness, right arm thrombosis, sinovenous thrombosis
- (25) NA = Not Applicable (no history of thrombosis)



Protein S Deficiency And Thrombophilia



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[06] What medication has this person taken? Please list all the drugs used for treatment or prevention of thrombosis (e.g. warfarin) even if you are not taking them now. Do not include drugs used to treat other conditions even if you feel they are for an associated condition (e.g. anti-depressants).

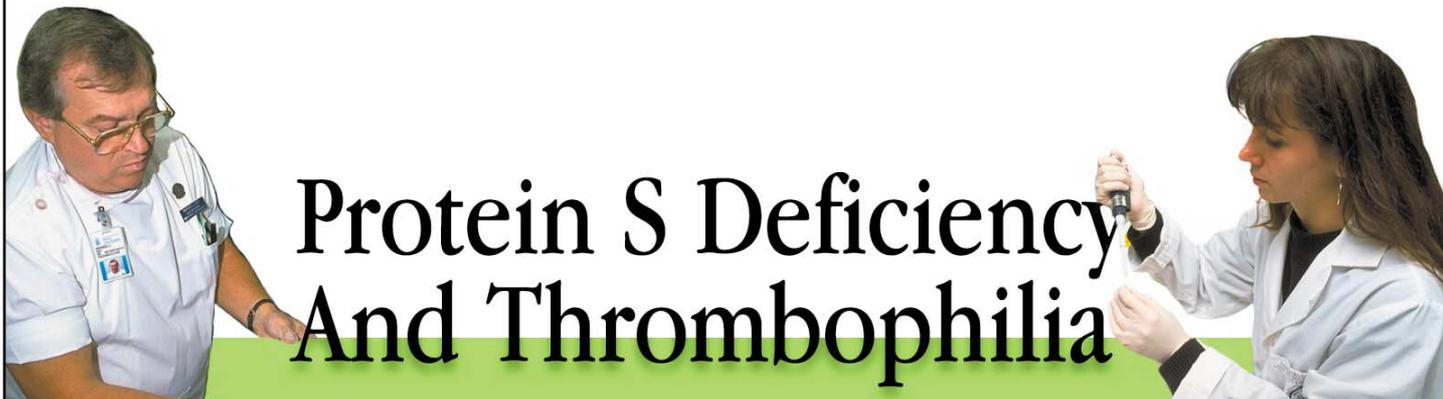
- (62) W/C = Coumadin or Warfarin (11 Both Coumadin and Warfarin)
- (39) W = Warfarin
- (34) C = Coumadin
- (26) LMH or Lovenox or Fragmin
- (24) H = Heparin
- (19) A = Aspirin
- (15) LMH = Low Molecular Heparin (e.g. Fragmin, Lovenox)
- (15) L = Lovenox
- (1) F = Fragmin
- (3) Enter the name of any drug if it is not shown: marcumar, plavix
- (13) NA = Not Applicable (no history or risk of thrombosis)

[07] How healthy is this person? Some people might suffer from several health conditions such as hayfever, asthma, allergies, frequent headaches, etc. We want to find out your opinion.

- (1) don't know or don't want to answer
- (25) very healthy
- (29) usually healthy
- (21) just average
- (10) frequently unhealthy
- (1) very unhealthy

[08] How good is the lifestyle of this person? Some people find it difficult to cope with poor health and another person can take it in their stride. A range of factors can play a part in this, such as living on your own, being disabled, on low income or perhaps living in a remote area. We want to find out your opinion.

- (0) don't know or don't want to answer
- (62) a good quality of life
- (14) able to cope without assistance
- (10) needs help from time to time
- (1) needs regular help
- (0) unable to cope



Protein S Deficiency And Thrombophilia

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[09] What topics are important to PSD Research? We want to ensure that research is relevant to our needs. If you can think of a topic that PSD Research should focus on please suggest it here. You are encouraged to list anything you think is important (e.g. medication). Some topics may be beyond the capability of an online survey but are still relevant as it may be possible for your suggestion to be followed up in some other way.

Many comments were included with the survey responses. These are summarised below. There was strong interest in gene therapy, effects of anti-coagulants especially during pregnancy, raising awareness of risk factors and the hereditary nature of Protein S Deficiency.

Medication: pregnancy, long term effects of anti-coagulants and side effects, alternative medicine, synthetic Protein S, home testing, safety of medication, interactions

Genetics: gene therapy, age of testing children, can PSD skip generations, genetic links and interactions with auto-immune diseases, prevention of passing PSD to children, diagnosis and screening

Education: prevention, doctor training, related conditions, exercise, diet, effects of altitude on INR, liver function, how children are affected, risk factors, age onset of risk factors, surgical risks, life expectancy

Lifestyle: pain control, feeling cold, effects of passive smoking, obtaining insurance, wound healing, blood circulation, abdominal thrombosis, effect on artery shrinkage, suitability of HRT, suitability of IVC filter, heart surgery

[10] Technical Information

(87) Anonymous

Details were automatically created by the web server and recorded in case it became necessary to solve technical issues.



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Discussion

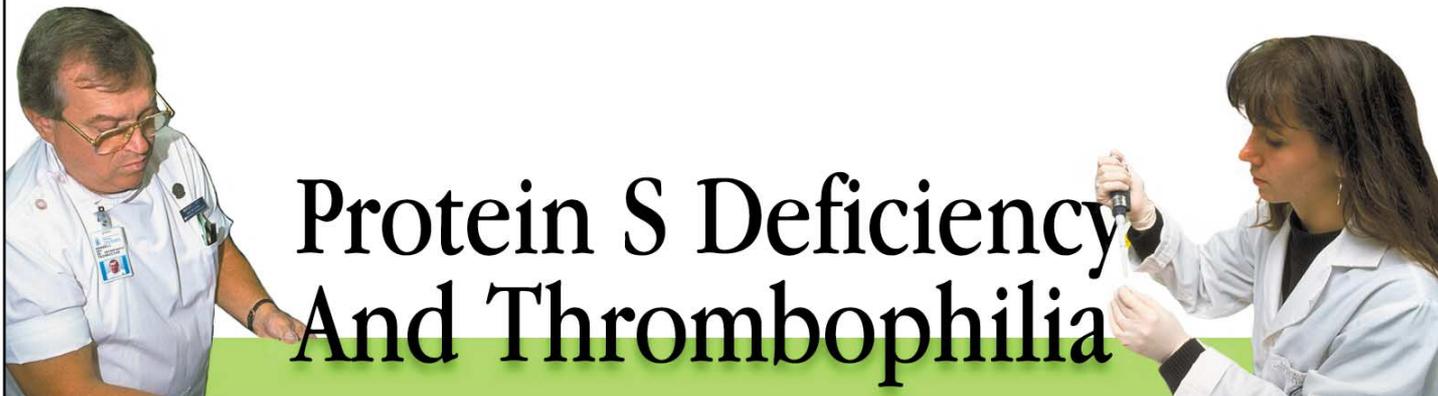
We expect that the survey results will be of interest to professionals and non-professionals alike. The survey design was kept open ended and flexible so that if patterns emerged in the results then maybe they could be put into context. For example there were nearly twice as many female respondents than male and this reflects the make-up of our forum membership and the general Internet trend for women to be the most active on health related web sites.

However some of the results are rather unexpected and might be misleading. On face value the results suggest most people feel healthy and have a good quality of life and therefore don't have any serious issues to speak of. In our online forum we know that this is not the case. Perhaps those that are coping better are more likely to engage in an online survey but it is probably also true that dealing with a life threatening condition can invoke a fighting spirit and a sense of optimism. It is felt the positive impression that has been given is indicative of people that have been through adversity and now want to make the most out of life and are looking forward to a better future.

A very large number of people said they were diagnosed with PSD after an actual or suspected thrombosis, especially hereditary PSD which is rather unfortunate. This suggests that screening for hereditary PSD is inadequate. These people are only learning of their genetic predisposition after becoming ill and the opportunity to make lifestyle changes to reduce other risk factors (such as smoking), taking medication as a preventative measure or being alert to symptoms is being missed out upon. The feasibility of a screening programme for hereditary PSD might be played down because modern treatment techniques for thrombosis have significantly reduced the mortality rate but this doesn't take into account the avoidable loss of quality of life and non-recoverable aspects such as Post Thrombotic Syndrome.

We noted that all of the people (27) reporting Pulmonary Embolism (PE) also reported Deep Vein Thrombosis (DVT) so nobody reported PE without DVT. It is customary for people with their first thrombosis (usually DVT) to stay on anti-coagulants for just three to six months but it might be worth investigating whether people that have been diagnosed with PSD should consider indefinite anti-coagulant therapy following a first thrombosis especially when considering the significant mortality rate of PE.

It is interesting to note that none of the people reporting PE (27) also reported stroke, and none of the people reporting stroke (8) also reported PE which suggests that thrombophiliacs might be to susceptible to one or the other but not both PE and stroke.



Protein S Deficiency And Thrombophilia

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Of the people reporting no thrombosis (25) we found that (24) had Protein S Deficiency. Of these we found that (8) have taken Coumadin, (4) have taken Aspirin, (5) have taken Heparin or Low Molecular Heparin (such as Lovenox) and more than half (13) no medication. It is unclear why there is such a wide variation in the prescribed medications being taken by these (thrombosis free) people but it is indicative of the comments in our forum regarding the dilemma of people who have been diagnosed with Hereditary PSD, perhaps as a result of family screening, working out whether they need to take medication.

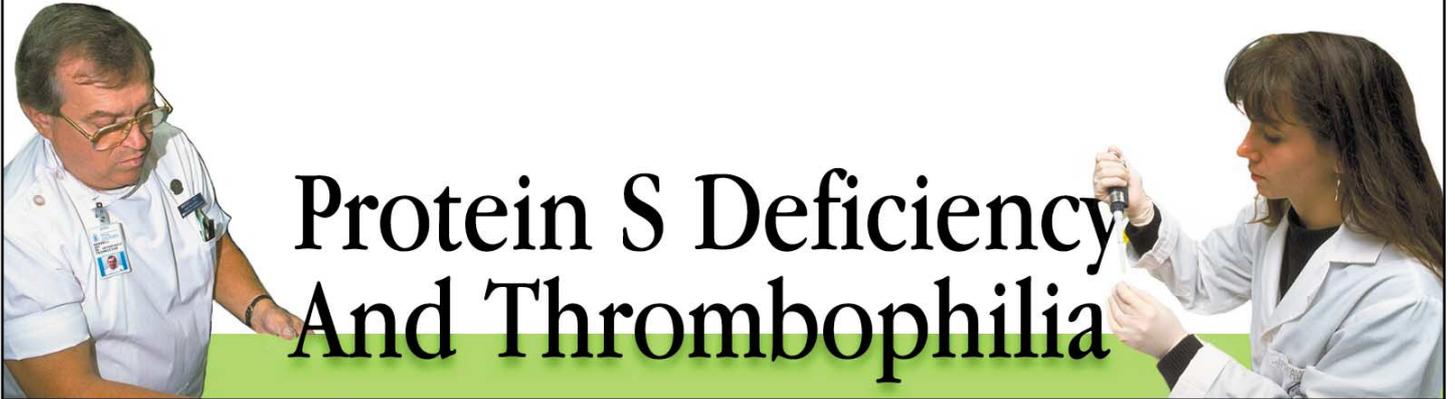
Conclusion

When we invited people to participate in our first survey we set out three main aims so that people knew their input would be valued and we could measure the success (or failure) of our initiative.

Our first aim was to demonstrate our capability to support research by describing health, lifestyle and experiences. We were pleased to see the strong support shown by everybody and such a high number of responses. We feel our group members have clearly indicated a readiness to provide information about themselves and would be prepared to participate in further surveys about Protein S Deficiency.

Our second aim was to gather some meaningful information that could be easily interpreted and used to produce a report. The structure and nature of questions were organised so it was possible to initially validate who the information was about and then describe qualitative results. Quantitative information would have been of little value because the selection criteria for survey candidates was biased in many ways due to the collection method (non-clinical and Internet based and without a control group).

Our third aim was to learn how to improve the process of gathering similar information and identify areas of special interest for following up. Two people reported technical problems in viewing the online form but a workaround was found and they submitted results in the normal way. Some of the responses (such as medication used) were answered in different ways making analysis more complicated. However it would be difficult to collect this type of information in any other way due to the variety of generic drugs and the various brand names used around the world. The additional comments received were considered very useful and will undoubtedly steer the direction of future surveys and website content.



Protein S Deficiency And Thrombophilia

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