

Left to Right -
Shelley S, Tom A, James K,
David P, Riyad E, Tom B,
Tina M, Jacques H, Silvia M,
Bruce R, Jeanne B, Gord S



Network of Rare Blood Disorder Organizations First Meeting

June 19 & 20, 2004
Toronto, Ontario



James, Corrado & Riyad



NRBDO Fall Conference 2017

Comprehensive Care

Bruce Ritchie MD, FRCPC³

Director VTE Clinic, UofA Hospital

Director Comprehensive Bleeding Disorder Clinic

Director Comprehensive Rare Blood Disorder Clinic

Director Canadian BioSample Repository (CBSR)

Nov-2017

Conflicts of Interest

- Principle Investigator in Trials: VTE, HAE, SCIg, Chelation, Porphyria,
- **Unpaid** consulting advice/Ad boards for: Alnylam
Baxter/Baxalta/Shire, Bayer, Boehringer-Ingelheim, Bristol-Myers-Squibb, Canadian Blood Service, Canadian Hemophilia Society, Covidien, CSL Behring, Dyax, Genzyme, Leo Pharmaceuticals, Novartis, Novo Nordisk, Octapharma, Pharming, Pfizer, Servier, Sanofi, Talecris, Wyeth
- In lieu of Honoraria for all activities, I request donations be made to the University of Alberta for summer students
Received funding for Investigator Initiated studies (biobanking) from: Baxter, CSL Behring, Genzyme, Leo, Novartis, Novartis, Novo Nordisk, Pfizer, Shire (no), Wyeth.
- I prepared all slides myself
- I may talk about off-label use of medication

NRBDO Fall Conference 2017

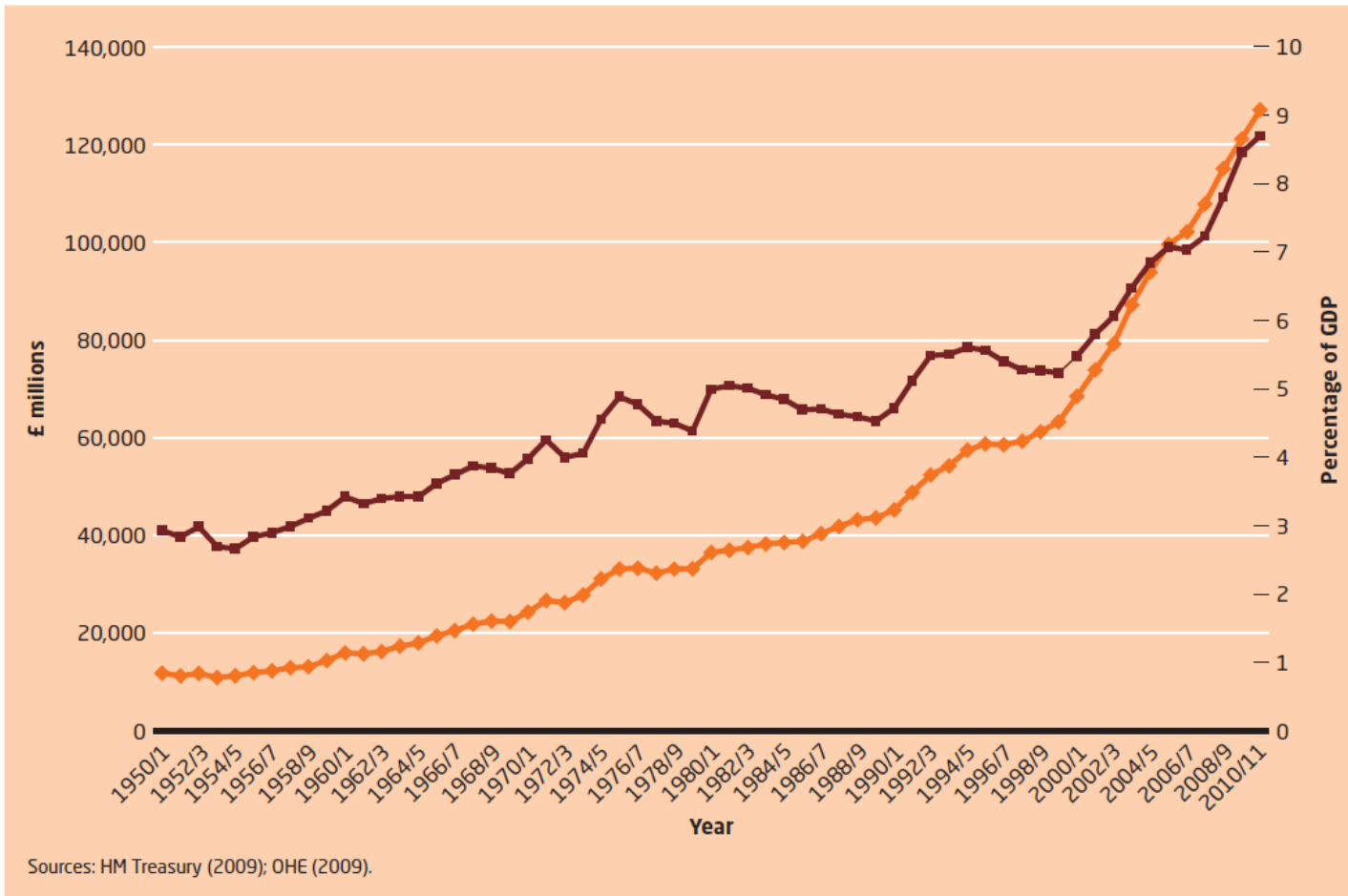
Comprehensive Care

1. Economic issues/Technology & Productivity
2. Comprehensive Care/Rare Diseases
3. Alberta Comprehensive Care Programs
4. Canada Health Care
5. PharmacoEconomic data

Nov-2017

Costs of UK National Health System

Figure 1 Real net spending on the UK NHS at 2010/11 prices and as a percentage of GDP

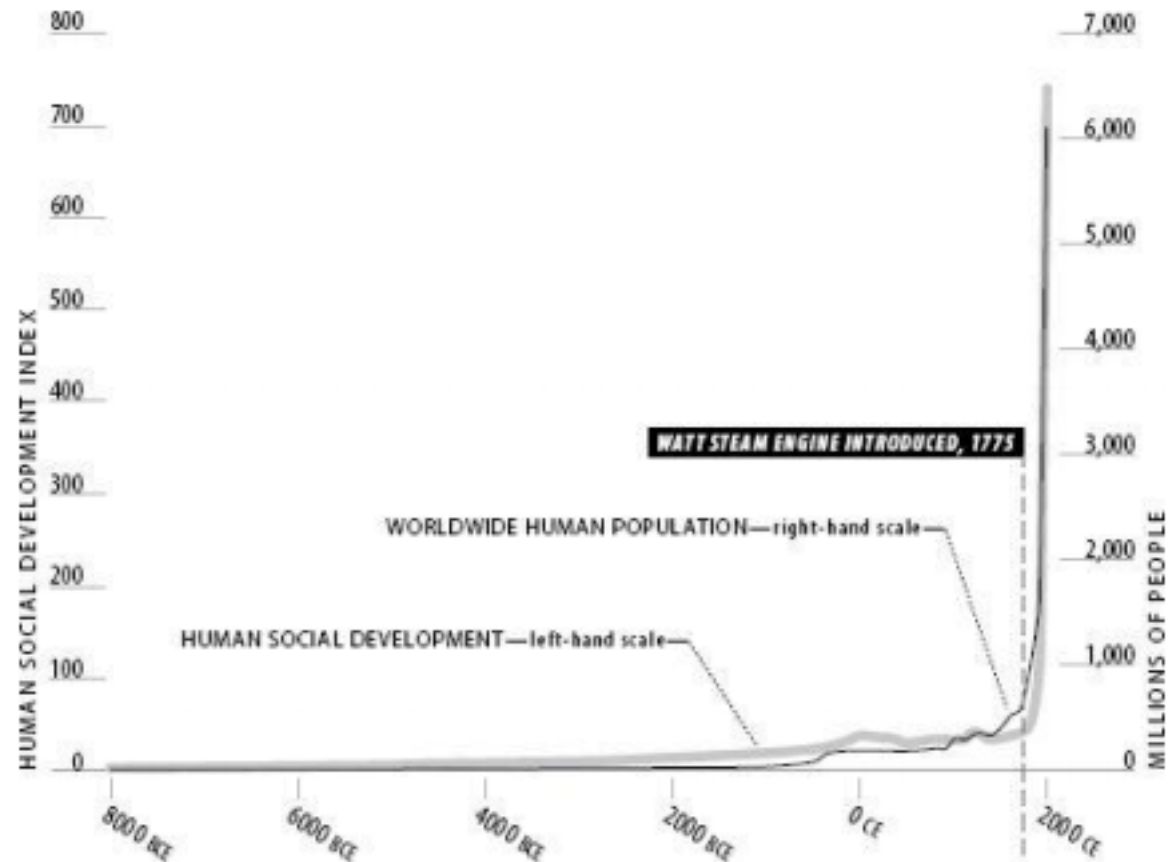
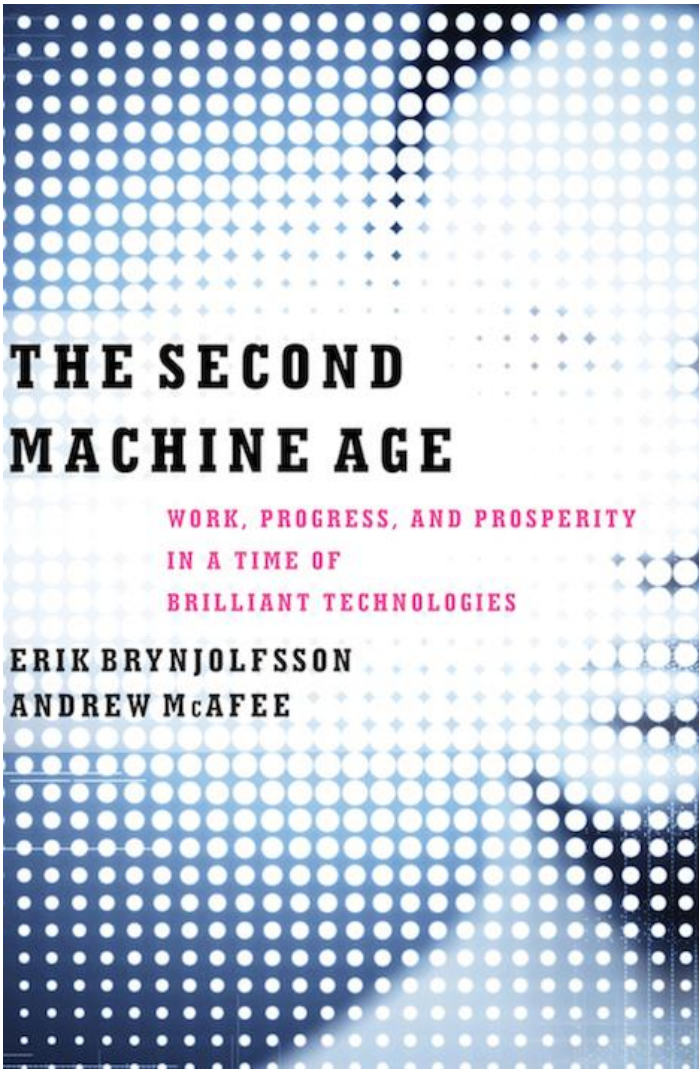


Real spending (left axis)
Per cent GDP (right axis)

The Second Machine Age

Work, Progress, and Prosperity in a time of Brilliant Technologies

<http://www.secondmachineage.com/>



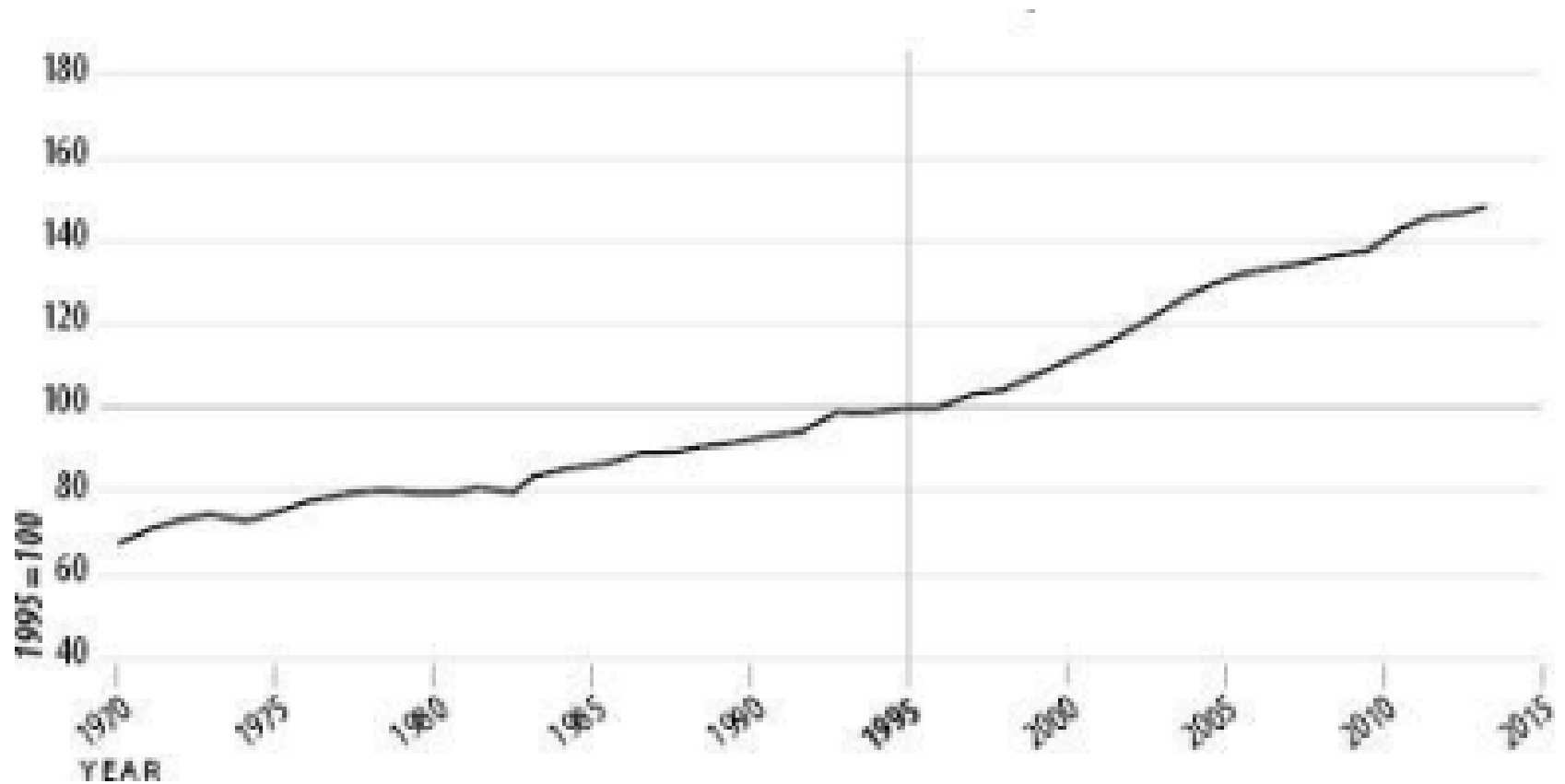
Some economic thoughts

Most economic fallacies derive from the tendency to assume that there is a fixed pie, that one party can gain only at the expense of another.

Milton Friedman 32-Jul-1912 – 16-Nov-2006

Some economic thoughts

FIGURE 7.1 Labor Productivity



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Nov-2017

Centre for Bleeding Disorders/Rare Blood Disorder Program Alberta Health Services

Comprehensive Care/Home Treatment Programs in:

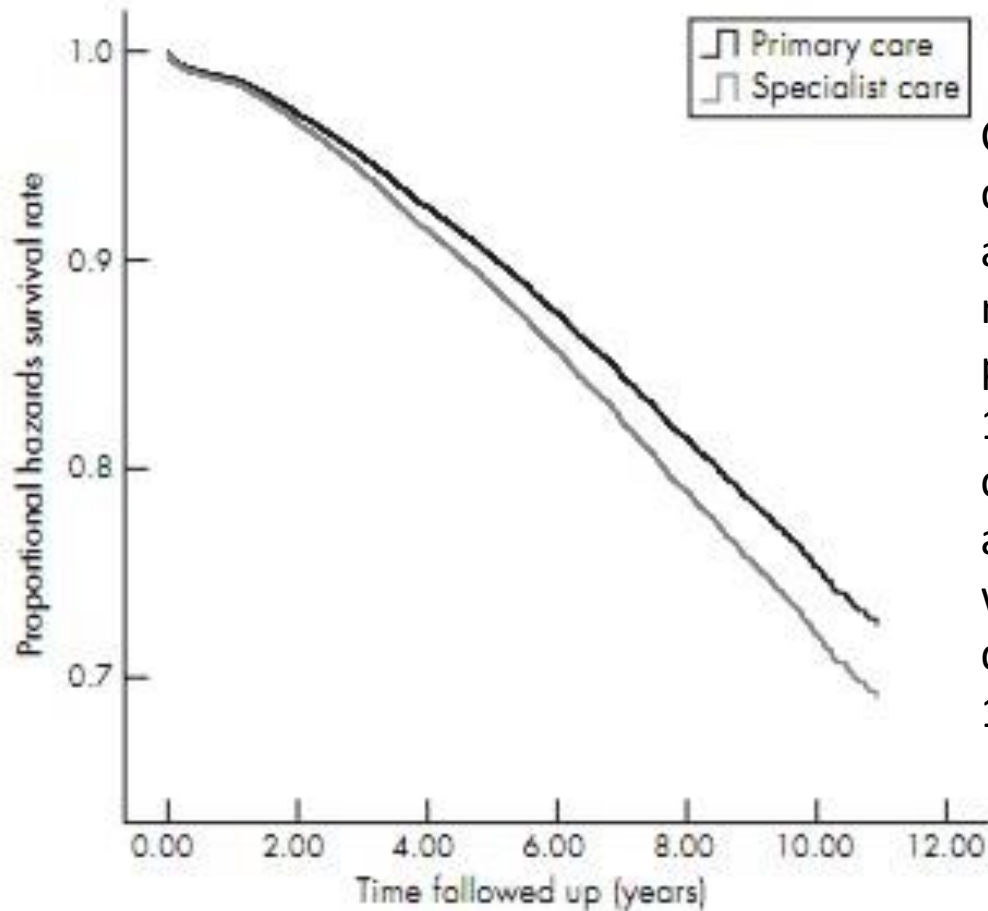
1. **Bleeding Disorders:** Nurse, Social Work, Physio, Orthopedics, Dentist, Hematologist,
2. **HAE:** Social Work, Nurse, Physicians (Heme, Allergy)
3. **Immunodeficiency:** Social Work, Nurse, Physicians (Heme, Immunology, Rheumatology, Respiratory Med, General Med)
4. **Hemoglobinopathy:** Nurse, Social Work, Physicians 3X Heme)
5. **Porphyria:** Social Worker, Nurse, Hematologist
6. **Ehlers Danlos syndrome:** Social Worker, Nurse, Hematologist, General Med-Hypertension)
7. **+/- Gaucher:** Nurse, Social Worker,

Centre for Bleeding Disorders/Rare Blood Disorder Program Alberta Health Services

Policies:

1. **Annual Visit:** Appointment for next year at end of visit
2. **Treatment Logs/CBDR:** Required to stay on Home Treatment for product tracking for recall, for justification of Rx - breakthrough
3. **Testing:** ie Immunoglobulin levels/CBC/retic (SCIG); CBCd (Hydroxyurea), ferritin, iron saturation (Chelation),
4. **Support: 8:00-4:00** Social Worker, Nurse
5. **Support: 24/7 access:** On-call, ER protocols, “Factor First” cards
6. **Family Physician: !!!!!**

The importance/impotence of specialty care: strategies for “normalizing” survival



Compared with patients seen by primary care doctors alone, patients seen by specialists and primary care doctors were more likely to receive recommended treatments (all $p(0.001)$), but were more likely to die (13.1% v 11.7%, adjusted hazard ratio (HR) 1.17, 95% confidence interval (CI) 1.08 to 1.27). This association persisted even in patients without co-morbidities or target organ damage (adjusted HR 1.16, 95% CI 1.01 to 1.34).

Figure 2 Kaplan-Meier graph of survival in patients treated by specialists or primary care doctors (crude mortality).

NRBDO Fall Conference 2017

Comprehensive Care

1. Economic issues/Technology & Productivity
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Nov-2017

A COMPARISON OF THE PATIENT-BORNE COSTS OF THERAPY WITH GAMMA GLOBULIN GIVEN AT THE HOSPITAL OR AT HOME

Ann Gardulf

Huddinge University Hospital

Gudrun Möller

Egon Jonsson

The Swedish Council on Technology Assessment in Health Care

30 patients with CVID or XLA

26-60 ml at 10-20 ml/hr

Products equivalent (free)

\$265/yr for travel, parking vs \$40/yr for syringes, needles etc

Slight benefit in time requirements

Int J Technology Assess in Health Care, 1995;11:345-53

Table 1. Resource use for base case models

| Resources | SCIG | IVIG |
|-------------------------|------|------|
| First year | | |
| Infusion supplies (day) | 208 | |
| RN (h) | – | |
| RN/manager (h) | 12 | |
| Unit clerk (h) | – | |
| Scheduling clerk (h) | – | |
| Ward aid (h) | – | |
| Second and third years | | |
| Infusion supplies (day) | 208 | |
| RN (h) | – | |
| RN/manager (h) | 6 | |
| Unit clerk (h) | – | |
| Scheduling clerk (h) | – | |
| Ward aid (h) | – | |

| Parameter | Unit cost (Can\$) | Sources |
|--|--------------------|--|
| Supplies | | |
| SCIG infusion set | 1.15 | Ministry of Health, Product Distribution Centre (2011) |
| Winged needle butterfly (4 times/week) | | |
| IVIG infusion (total supplies) | 13.54 | Ministry of Health, Product Distribution Centre (2011) |
| Personnel (h) | | |
| RN/manager (SCIG treatment) | 52.5 | British Columbia Nurses' Union (2011) |
| RN (IVIG treatment) | 35.00 ¹ | British Columbia Nurses' Union (2011) |
| Unit clerk | 19.86 | Hospital Employees' Union (2011) |
| Scheduling clerk | 20.77 | Hospital Employees' Union (2011) |
| Ward aid | 19.86 | Hospital Employees' Union (2011) |

Martin A, Lavoie L, Goetghebeur M, Schellenberg R. Economic benefits of subcutaneous rapid push versus intravenous immunoglobulin infusion therapy in adult patients with primary immune deficiency. *Transfus Med.* 23(1):55-60, 2013 Feb.

Table 3. Cost-minimisation model results (base case)

| Costs per patient | SCIG (Can\$) | IVIG (Can\$) | Increment: SCIG – IVIG (Can\$) |
|---|-----------------|-----------------|--------------------------------------|
| First year | | | |
| Infusion supplies | 239 | 194 | 46 |
| RN or RN/manager salary | 630 | 2002 | -1372 |
| Other personnel salary | | | |
| Unit clerk | - | 114 | |
| Scheduling clerk | - | 149 | |
| Ward aid | | 114 | |
| Total personnel salary | 630 | 2378 | -1748 |
| Total (first year) | 869 | 2571 | -1702 |
| Second year | | | |
| Infusion supplies | 239 | 194 | 46 |
| RN or RN/manager salary | 315 | 2002 | -1687 |
| Other personnel salary | | | |
| Unit clerk | - | 114 | |
| Scheduling clerk | - | 149 | |
| Ward aid | - | 114 | |
| Total personnel salary | 315 | 2378 | -2063 |
| Total (second year) | 554 | 2571 | -2017 |
| Third year | | | |
| Infusion supplies | 239 | 194 | 46 |
| RN or RN/manager salary | 315 | 2002 | -1687 |
| Other personnel salary | | | |
| Unit clerk | - | 114 | |
| Scheduling clerk | - | 149 | |
| Ward aid | - | 114 | |
| Total personnel salary | 315 | 2378 | -2063 |
| Total (third year) | 554 | 2571 | -2017 |
| Total cost per patient (3 years) | 1978 | 7714 | -5736 |

Martin A, Lavoie L, Goetghebeur M, Schellenberg R. Economic benefits of subcutaneous rapid push versus intravenous immunoglobulin infusion therapy in adult patients with primary immune deficiency. *Transfus Med.* 23(1):55-60, 2013 Feb.

TABLE I. Assessment of time per year and per patient spent by parents and care providers according to treatment

| | IV, mean \pm SD | SC, mean \pm SD | SC-Ig minus IV-Ig, mean (95% CI) | P value (signed rank test) |
|--|-------------------|-------------------|-------------------------------------|-------------------------------|
| Time (h) spent by parents (treatment supervision and administration and time spent in hospital visits) | 160 \pm 105 | 78 \pm 40 | -82 (-123 to -40) | <.001 |
| Nurse (h) | 60 | 7.5 | -52.5 | |
| Technician (h) | 3 | 1 | -2 | |
| Administrative assistant (h) | 3 | 1 1/3 | -1 2/3 | |

TABLE II. Medical and nonmedical resource use and costs over the first year by dollars per patient

| | IV, mean \pm SD | SC, mean \pm SD | SC-Ig minus IV-Ig, mean (95% CI) | P value (signed rank test) |
|--|---------------------|--------------------|-------------------------------------|-------------------------------|
| Nonmedical resources | | | | |
| (1) Travel expenses | | | | |
| Transports (taxi, bus, metro, own car) | 773 \pm 921 | 193 \pm 230 | -580 (-865 to -295) | |
| Hotel | 182 \pm 476 | 46 \pm 119 | -137 (-284 to 11) | |
| Parking | 63 \pm 73 | 16 \pm 18 | -48 (-70 to -25) | |
| Total | 1,019 \pm 1,222 | 255 \pm 305 | -764 (-1,142 to -386) | <.001 |
| (2) Time spent by parents (treatment supervision and administration and time-spent in hospital visits) | 3,687 \pm 2,430 | 1,807 \pm 935 | -1,880 (-2,840 to -920) | <.001 |
| (3) Other resources (Emla cream) | <1 | 69 \pm 126 | 69 (17 to 121) | .03 |
| Total nonmedical resources (1) + (2) + (3) | 4,706 \pm 3,109 | 2,131 \pm 976 | -2,575 (-3,773 to -1,378) | <.001 |
| Medical resources | | | | |
| (4) Nurse, technician, administration (SJH) | 1,867 | 273 | -1,594 | |
| (5) Pump, infusion materials | 136 | 780 | 644 | |
| (6) Medical services (consultations) | 2,685 \pm 5,508 | 1,918 \pm 2,882 | -767 (-2,853 to 1,318) | .14 |
| (7) Pharmaceuticals | 1,367 \pm 3,630 | 1,387 \pm 3,164 | 20 (-1,024 to 1,063) | .81 |
| Total medical resources (4) + (5) + (6) + (7) | 6,055 \pm 6,197 | 4,358 \pm 4,495 | -1,698 (-3,242 to -153) | <.001 |
| Total medical and nonmedical resources | 10,761 \pm 7,874 | 6,488 \pm 4,571 | -4,273 (-6,215 to -2,331) | <.001 |
| Immunoglobulins | 13,084 \pm 9,225 | 12,556 \pm 8,046 | -528 (-1,716 to 660) | .36 |
| Total medical and nonmedical resources + immunoglobulins | 23,845 \pm 13,245 | 19,044 \pm 9,476 | -4,801 (-7,419 to -2,183) | <.001 |
| Cost according to perspective | | | | |
| Patient perspective (1) + (2) + (3) + (5) (only for SC) | 4,706 \pm 3,109 | 2,911 \pm 977 | -1,795 (-2,993 to -598) | .05* |
| Government perspective (4) + (5) (only for IV) + (6) + (7) | 6,055 \pm 6,197 | 3,578 \pm 4,495 | -2,478 (-4,022 to -933) | .003 |

Ducruet, Thierry. Levasseur, Marie-Claude. Des Roches, Anne. Katal, Ayman. Dicaire, Renee. Haddad, Elie.

Pharmacoeconomic advantages of subcutaneous versus intravenous immunoglobulin treatment in a Canadian pediatric center. *J Allergy Clin Immunol.* 131(2):585-7.e1-3, 2013 Feb.



SHORT REPORT

Open Access

Implications to payers of switch from hospital-based intravenous immunoglobulin to home-based subcutaneous immunoglobulin therapy in patients with primary and secondary immunodeficiencies in Canada

William C Gerth^{1*}, Stephen D Betschel² and Arthur S Zbrozek³

Gerth WC, Betschel SD, Zbrozek AS. Implications to payers of switch from hospital-based intravenous immunoglobulin to home-based subcutaneous immunoglobulin therapy in patients with primary and secondary immunodeficiencies in Canada. *Allergy, Asthma & Clinical Immunology* 2014, **10**:23. <http://www.aacijournal.com/content/10/1/23>

Implications to payers of switch from hospital-based intravenous immunoglobulin to home-based subcutaneous immunoglobulin therapy in patients with primary and secondary immunodeficiencies in Canada

Table 3 Estimated number of IgG-treated patients with PID and SID in Canada by province

| Province | 2012 population (Thousands) | PID | SID | PID/SID combined |
|----------------------------|--------------------------------|----------------|----------------|---------------------|
| Alberta | 3,873.7 | 396.1 | 213.3 | 609.3 |
| British Columbia | 4,622.6 | 472.6 | 254.5 | 727.1 |
| Quebec | 8,054.8 | 823.6 | 443.5 | 1,267.1 |
| Manitoba | 1,267.0 | 129.5 | 69.8 | 199.3 |
| New Brunswick | 756.0 | 77.3 | 41.6 | 118.9 |
| Newfoundland & Labrador | 512.7 | 52.4 | 28.2 | 80.7 |
| Nova Scotia | 948.7 | 97.0 | 52.2 | 149.2 |
| Ontario | 13,505.9 | 1,380.9 | 743.6 | 2,124.5 |
| Prince Edward Island | 146.1 | 14.9 | 8.0 | 23.0 |
| Saskatchewan | 1,080.0 | 110.4 | 59.5 | 169.9 |
| Territories | 113.1 | 11.6 | 6.2 | 17.8 |
| Total | 34,880.6 | 3,566.0 | 1,920.0 | 5,486.0 |

Gerth WC, Betschel SD, Zbrozek AS. Implications to payers of switch from hospital-based intravenous immunoglobulin to home-based subcutaneous immunoglobulin therapy in patients with primary and secondary immunodeficiencies in Canada. *Allergy, Asthma & Clinical Immunology* 2014, 10:23. <http://www.aacijournal.com/content/10/1/23>

Implications to payers of switch from hospital-based intravenous immunoglobulin to home-based subcutaneous immunoglobulin therapy in patients with primary and secondary immunodeficiencies in Canada

Table 1 Annual net economic benefit in Canada: Nurse time and compensation per patient switched from hospital-based IVIg to home-based SCIg

| | Year 1 |
|---|--------|
| Home-based SCIg Therapy | |
| Nurse hours: training | 6 |
| Nurse hours: monitoring | 6 |
| Nurse compensation, C\$ | 691 |
| Hospital-based IVIg therapy | |
| Nurse Hours: IV administration and monitoring | 57.2 |
| Nurse Compensation, C\$ | 3,294 |
| Net economic benefit (IVIg – SCIg) | |
| Savings: nurse hours | 45.2 |
| Savings: nurse compensation, C\$ | 2,603 |

Table 2 Annual net economic benefit in Canada: Nurse time and compensation per 37 patients switched from hospital IVIg to home SCIg to recoup 1 nurse FTE

| | Year 1 | Subsequent years |
|---|---------|------------------|
| Home-based SCIg therapy | | |
| Nurse hours: training | 222 | 0 |
| Nurse hours: monitoring | 222 | 222 |
| Nurse compensation, C\$ | 25,565 | 12,783 |
| Hospital-based IVIg therapy | | |
| Nurse hours: IV administration and monitoring | 2,116.4 | 2,116.4 |
| Nurse compensation, C\$ | 121,862 | 121,862 |
| Net economic benefit (IVIg – SCIg) | | |
| Savings: nurse hours | 1,672.4 | 1,894.4 |
| Savings: nurse compensation, C\$ | 96,297 | 109,080 |

Gerth WC, Betschel SD, Zbrozek AS. Implications to payers of switch from hospital-based intravenous immunoglobulin to home-based subcutaneous immunoglobulin therapy in patients with primary and secondary immunodeficiencies in Canada. *Allergy, Asthma & Clinical Immunology* 2014, 10:23. <http://www.aacijournal.com/content/10/1/23>

Implications to payers of switch from hospital-based intravenous immunoglobulin to home-based subcutaneous immunoglobulin therapy in patients with primary and secondary immunodeficiencies in Canada

Table 4 Estimated 3-year savings (Hours, FTEs, Costs) from switching 50% to 75% of treated patients with PID/SID from hospital IVIg to home SCIg in Canada by province

| Province | Savings with 50% switch rate | | | Savings with 75% switch rate | | |
|-------------------------|------------------------------|--------------|----------------------------|------------------------------|--------------|----------------------------|
| | Hours | FTEs | Labor costs (C\$ millions) | Hours | FTEs | Labor costs (C\$ millions) |
| Alberta | 44,969 | 24.8 | \$ 2.59 | 67,453 | 37.2 | \$ 3.88 |
| British Columbia | 53,662 | 29.6 | \$ 3.09 | 80,493 | 44.4 | \$ 4.64 |
| Quebec | 93,506 | 51.6 | \$ 5.38 | 140,258 | 77.4 | \$ 8.08 |
| Manitoba | 14,708 | 8.1 | \$ 0.85 | 22,062 | 12.2 | \$ 1.27 |
| New Brunswick | 8,776 | 4.8 | \$ 0.51 | 13,164 | 7.3 | \$ 0.76 |
| Newfoundland & Labrador | 5,952 | 3.3 | \$ 0.34 | 8,928 | 4.9 | \$ 0.51 |
| Nova Scotia | 11,013 | 6.1 | \$ 0.63 | 16,520 | 9.1 | \$ 0.95 |
| Ontario | 156,786 | 86.5 | \$ 9.03 | 235,179 | 129.7 | \$ 13.54 |
| Prince Edward Island | 1,696 | 0.9 | \$ 0.10 | 2,544 | 1.2 | \$ 0.15 |
| Saskatchewan | 12,537 | 6.9 | \$ 0.72 | 18,806 | 10.4 | \$ 1.08 |
| Territories | 1,313 | 0.7 | \$ 0.08 | 1,969 | 1.1 | \$ 0.11 |
| Total | 404,918 | 223.3 | \$ 23.30 | 607,376 | 335.0 | \$ 34.98 |

Gerth WC, Betschel SD, Zbrozek AS. Implications to payers of switch from hospital-based intravenous immunoglobulin to home-based subcutaneous immunoglobulin therapy in patients with primary and secondary immunodeficiencies in Canada. *Allergy, Asthma & Clinical Immunology* 2014, 10:23. <http://www.aacijournal.com/content/10/1/23>

Implications to payers of switch from hospital-based intravenous immunoglobulin to home-based subcutaneous immunoglobulin therapy in patients with primary and secondary immunodeficiencies in Canada

| Province (80%) | Capacity increase (hours/month) |
|----------------|---------------------------------|
| Alberta 500 | 2,200 |
| Ontario 1700 | 7,480 |
| Canada 4400 | 19,360 |

Gerth WC, Betschel SD, Zbrozek AS. Implications to payers of switch from hospital-based intravenous immunoglobulin to home-based subcutaneous immunoglobulin therapy in patients with primary and secondary immunodeficiencies in Canada. *Allergy, Asthma & Clinical Immunology* 2014, 10:23. <http://www.aacijournal.com/content/10/1/23>

Questions



NRBDO Fall Conference 2017

Comprehensive Care

1. Economic issues/Technology & Productivity
2. Comprehensive Care/Rare Diseases
3. Alberta Comprehensive Care Programs
4. Canada Health Care
5. PharmacoEconomic data
6. Benefits not currently collected as economic benefit

HAE Home Treatment

Original Paper

International Archives of
**Allergy and
Immunology**

Int Arch Allergy Immunol 2012;157:417–424
DOI: 10.1159/000329635

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Accepted after revision: May 23, 2011
Published online: November 25, 2011

Safety and Efficacy of Physician-Supervised Self-Managed C1 Inhibitor Replacement Therapy

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James Koziol^c Sandra C. Christiansen^{a, d} Bruce L. Zuraw^{a, e}

^aUniversity of California, San Diego, ^bUnited States Hereditary Angioedema Association, ^cThe Scripps Research Institute, ^dSouthern California Kaiser Permanente, and ^eSan Diego VA Medical Center, La Jolla, Calif., USA

Louanne M. Tourangeau^a Anthony J. Castaldo^b Donna K. Davis^b James Koziol^c Sandra C. Christiansen^{a, d} Bruce L. Zuraw^{a, e}.
Safety and Efficacy of Physician-Supervised Self-Managed C1 Inhibitor Replacement Therapy. Int Arch Allergy Immunol
2012;157:417–424

HAE Home Treatment

Table 2. Interval efficacy and safety/tolerability results

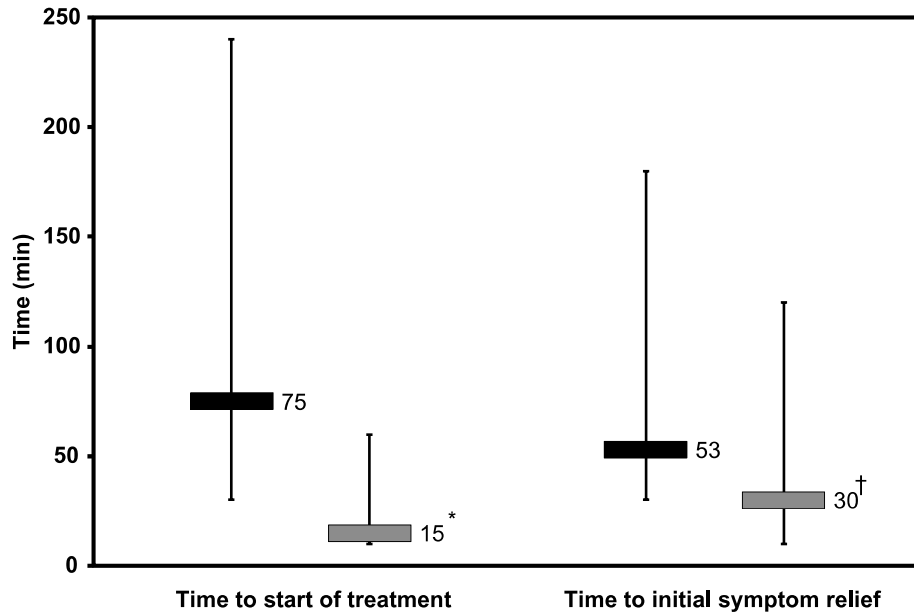
| Parameter | Clinic group (n = 18) | | Home group (n = 21) | | p value ¹ |
|---|-----------------------|--------------|---------------------|---------------|----------------------|
| | mean ± SD | median (IQR) | mean ± SD | median (IQR) | |
| Attack frequency/severity | | | | | |
| Average attacks/week | 0.83 ± 0.98 | 1 (0–2) | 1.02 ± 0.96 | 1 (0–2) | 0.82 |
| Attack severity (scale of 1–10) | 5.67 ± 2.69 | 5 (4–8) | 4.75 ± 2.46 | 4 (3–7) | <0.005 |
| Efficacy | | | | | |
| Attack duration, h | 34.2 ± 27.5 | 30.5 (12–48) | 8.0 ± 13.5 | 3.0 (1.5–6.0) | <0.0001 |
| Pain medications per week | 1.75 ± 3.13 | 0 (0–3) | 0.38 ± 0.98 | 0 (0–0) | <0.0001 |
| Safety/tolerability results | | | | | |
| Bleeding from injection site, % | 0.58 ± 7.6 | 0 (0–0) | 7.81 ± 26.9 | 0 (0–0) | <0.001 |
| Pain/inflammation at injection site, % | 1.73 ± 13.1 | 0 (0–0) | 7.21 ± 25.9 | 0 (0–0) | 0.009 |
| Infection at injection site, % | 0 ± 0 | 0 (0–0) | 0.30 ± 5.5 | 0 (0–0) | 0.47 |
| Difficulty with injection (scale of 0–3) | 0.03 ± 0.17 | 0 (0–0) | 0.31 ± 0.60 | 0 (0–0) | <0.0001 |
| Severity of C1INH side effects (scale of 0–3) | 0.22 ± 0.71 | 0 (0–0) | 0.01 ± 0.16 | 0 (0–0) | <0.0001 |

¹ Mann-Whitney rank-sum test.

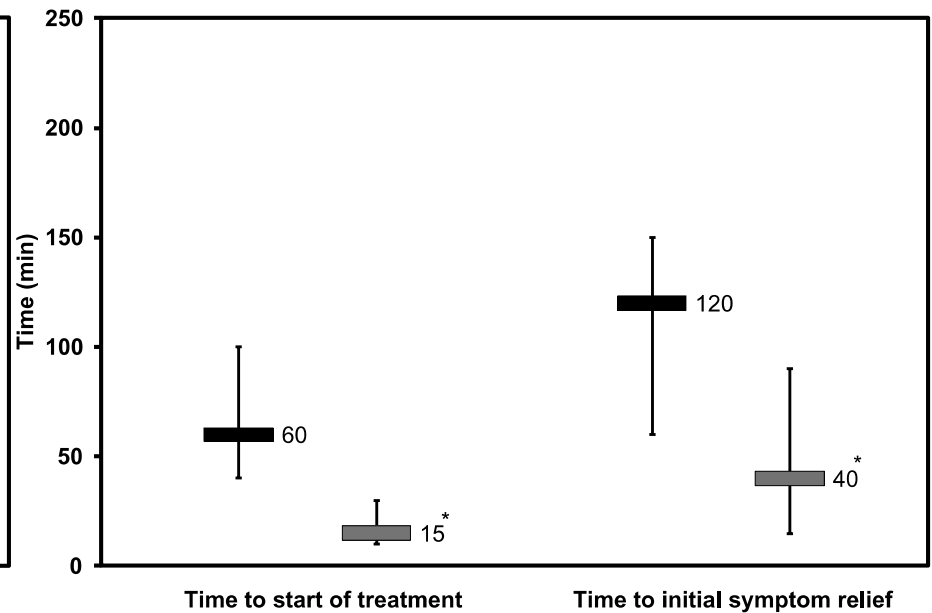
Home Therapy with Human C1-INH: Earlier Treatment and Trend Towards Higher Efficacy

Children and Adolescent Data Only

On Demand

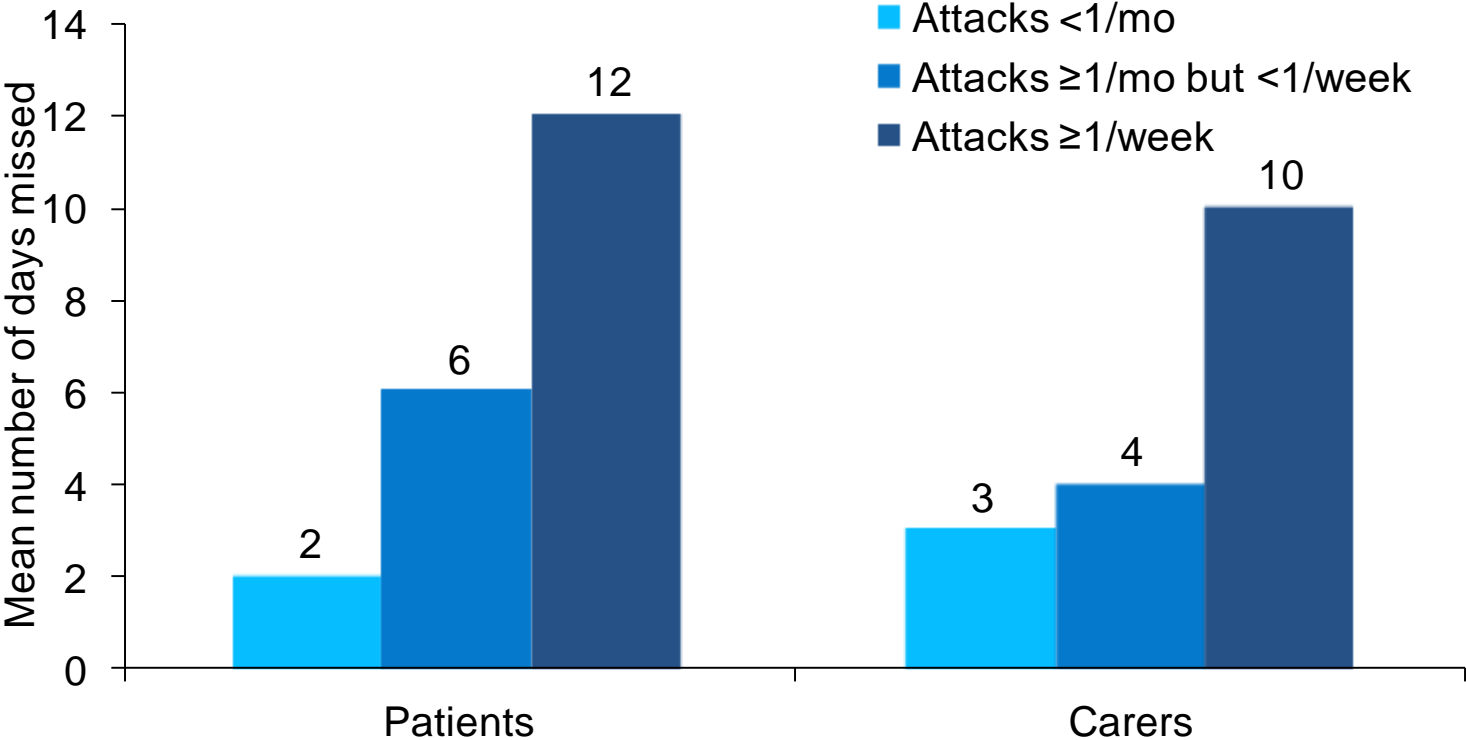


IRT



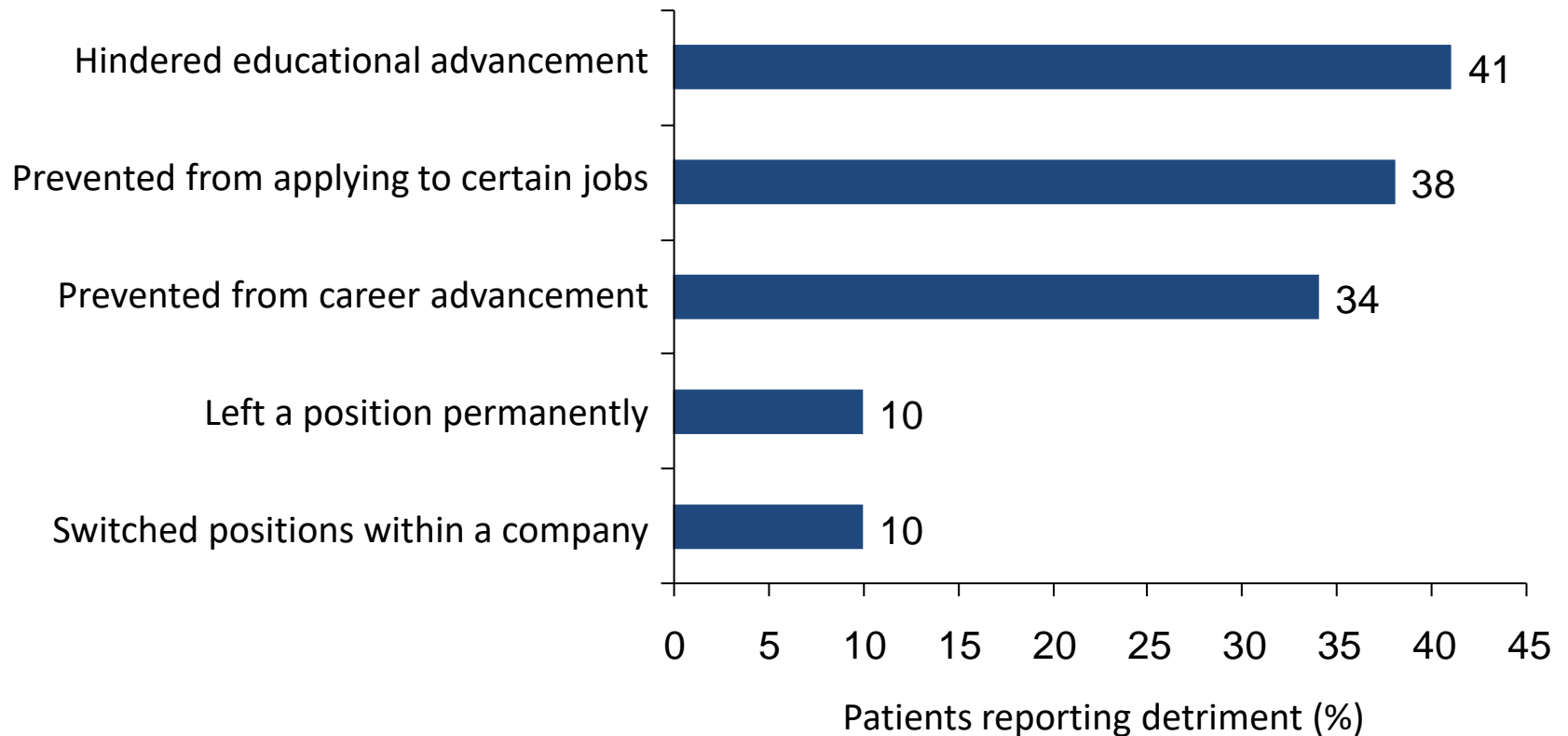
Absenteeism in between attacks over the past 6 months is associated with attack frequency

Mean number of days missed from work or school over the last 6 months by attack frequency



Note: For patients, includes time missed from work and/or school; for carers, includes time missed from work, school and/or leisure. Overall, 59% used carer help over the past 6 months.

Patients experience substantial detriments in career advancement due to HAE



Fatal laryngeal attacks and mortality in hereditary angioedema due to C1-INH deficiency

TABLE I. Characterization of 728 patients with HAE-C1-INH coming from 182 unrelated families

| | Undiagnosed (n = 201) | Diagnosed (n = 527) |
|--|--------------------------|------------------------|
| No. of patients | | |
| HAE-C1-INH type I | 184 | 498 |
| HAE-C1-INH type II | 17 | 29 |
| Patients who died | 201 | 13 |
| Patients who died due to HAE-C1-INH | 63 | 7 |
| Patients who died of other causes | 138 | 6 |
| Gender (M/F) | 110/91 | 229/298 |
| Age at the time of evaluation (y), mean ± SD | NA | 46.3 ± 19.5* |
| Age at death (y), mean ± SD | 62.5 ± 20.3 | 54.2 ± 23.3† |
| No. of families (n = 182) | | |
| HAE-C1-INH type I | 169 | |
| HAE-C1-INH type II | 13 | |
| ≥1 Death case due to HAE-C1-INH | 56 | |
| 1 Death case due to HAE-C1-INH | 44 | |
| 2 Death cases due to HAE-C1-INH | 10 | |
| 3 Death cases due to HAE-C1-INH | 1 | |
| 4 Death cases due to HAE-C1-INH | 1 | |

*Mean values of 514 living patients.

†Mean values of 13 patients.

Konrad Bork, MD,^a Jochen Hardt, PhD,^b and Gunther Witzke. Fatal laryngeal attacks and mortality in hereditary angioedema due to C1-INH deficiency. J Allergy Clin Immunol 2012;130:692-7

1000

the track is awesome

Im fine with breathing

like right now if i didnt
have the track

it would be bad cause

my throat is basically closed

Children and adults with primary antibody deficiencies gain quality of life by subcutaneous IgG self-infusions at home

Ann Gardulf, RN, PhD,^a Uwe Nicolay, Dipl Math,^a Oscar Asensio, MD,^b Ewa Bernatowska, MD, PhD,^c Andreas Böck, MD,^d Beatriz T. Costa-Carvalho, MD,^e Carl Granert, MD, PhD,^f Stefan Haag, MD, PhD,^g Dolores Hernández, MD, PhD,^h Peter Kiessling, PhD,^g Jan Kus, MD, PhD,ⁱ Nuria Matamoros, MD, PhD,^j Tim Niehues, MD, PhD,^k Sigune Schmidt, MD,^l Ilka Schulze, MD, PhD,^m and Michael Borte, MD, PhDⁿ
Stockholm, Sweden, Sabadell, Valencia, and Palma de Mallorca, Spain, Warsaw, Poland, Vienna, Austria, São Paulo, Brazil, and Marburg, Düsseldorf, Freiburg, Berlin, and Leipzig, Germany

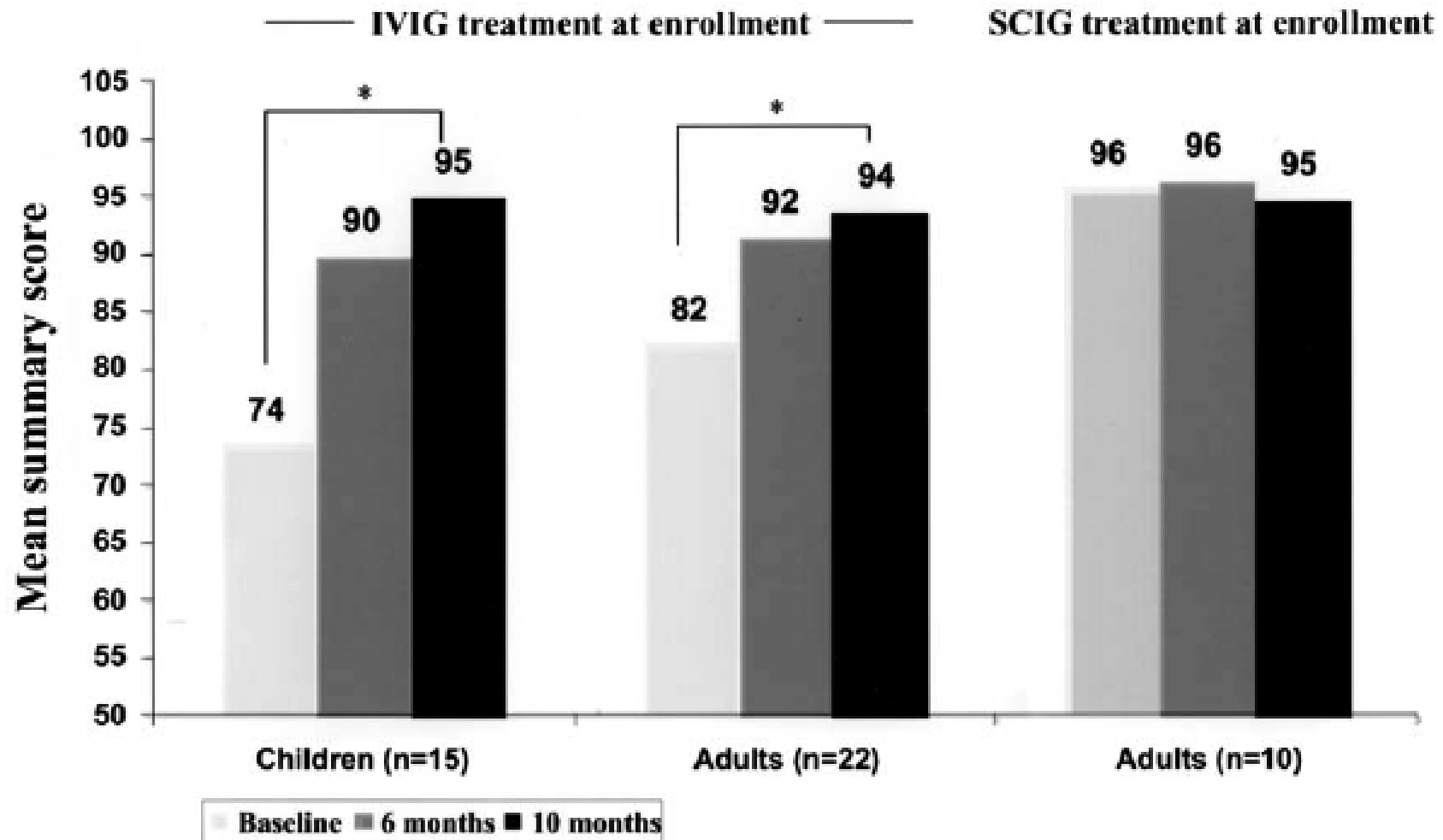
15 children (<14 y; IVIG therapy at enrollment); 32 adults (>14 y; 22 IVIG/10 SCIG).
Questionnaires at baseline, 6 & 10 months: Child Health Questionnaire–Parental Form 50 (children) or Short Form 36 (adults), Life Quality Index, and questions regarding therapy preferences. **Children reported:** better health, improved school/social functioning for children

Parents reported: Reduced emotional stress and limitations on personal time, fewer limitations on family activities

Adults reported: Improved vitality, mental health, and social functioning

Gardulf A, Nicolay U, Asensio O, Bernatowska E, Bock A, Costa-Carvalho B, Granert C, Haag S, Hernandez D, Kissling P, Kus J, Matamoros N, Niehues T, Schmidt S, Schulze I, Borte M. Children and adults with primary antibody deficiencies gain quality of life by subcutaneous IgG self-infusions at home. *J Allergy Clin Immunol*, 2004;114:936-42

Children and adults with primary antibody deficiencies gain quality of life by



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Krever Commission Report

On November 26, 1997, Health Minister Allan Rock, on behalf of the federal government, [released](#) the final report of the Commission of Inquiry on the Blood System in Canada (Krever Commission).

On November 25, 1998, the Health Minister observed the first anniversary of the report by [reaffirming the federal government's commitment to blood safety](#). He added that Health Canada [took action](#) concerning each one of the commission's final recommendations that concerned the Department.

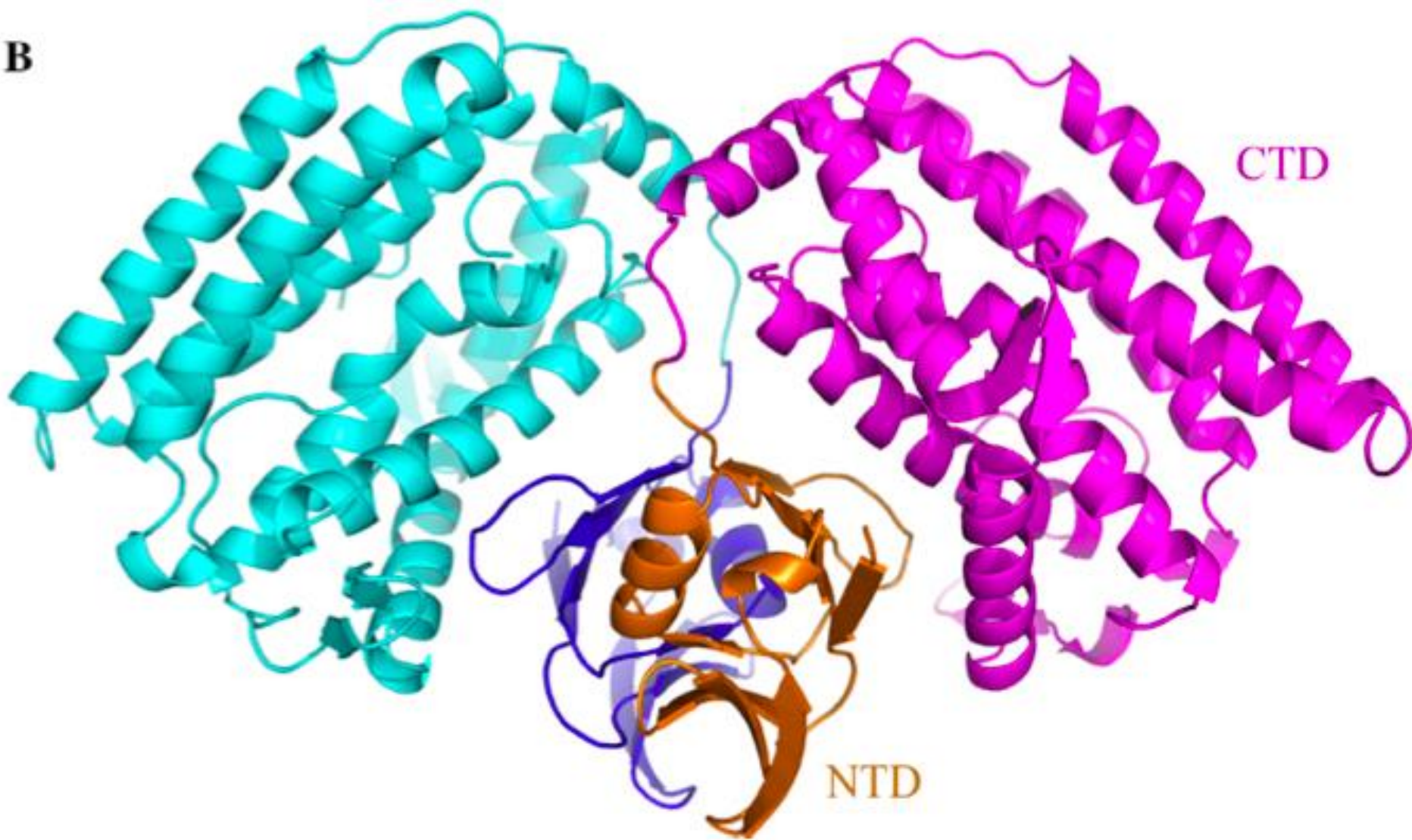
Report - [Table of Contents](#)

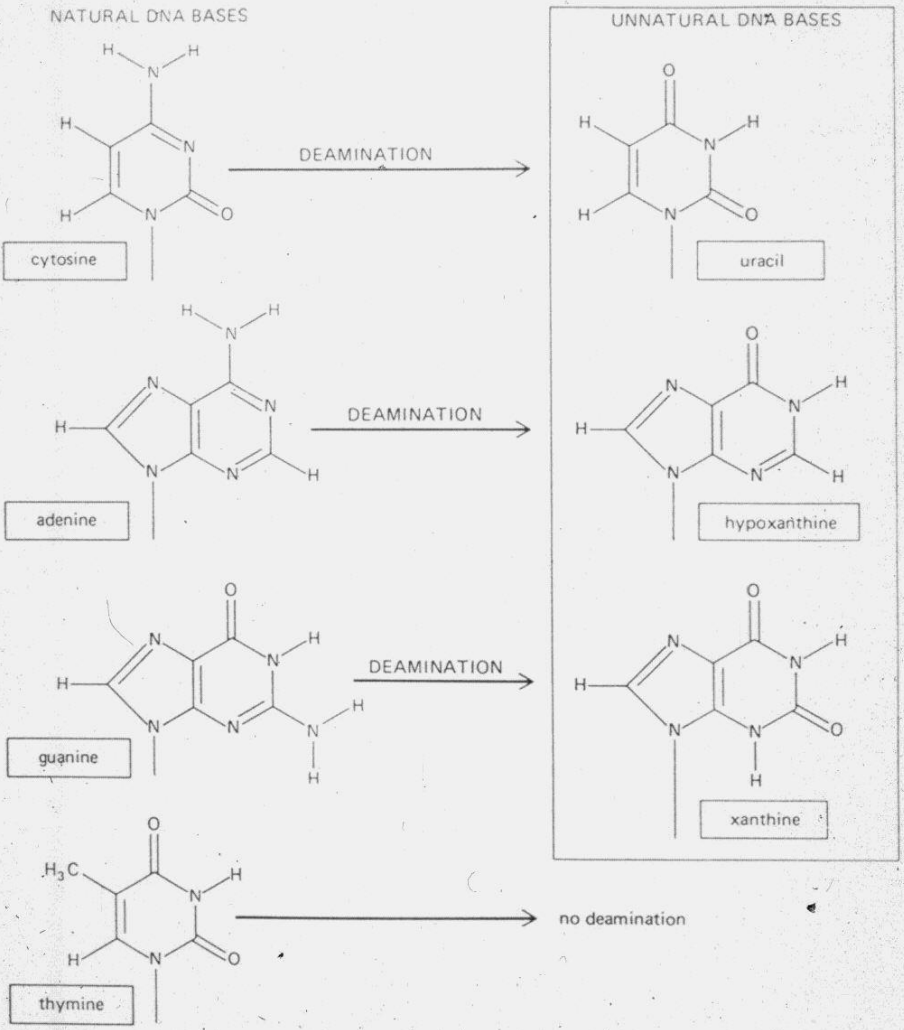
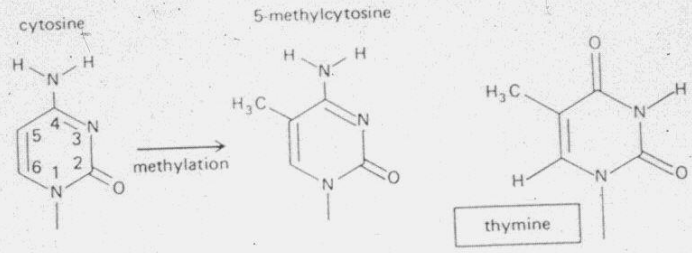
How to obtain the report

- View at [public libraries](#) participating in the Depository Services Program
- [Access PDF files](#) archived in the National Library of Canada Electronic Collection. (This requires free [Adobe Acrobat](#) software for viewing). The original printed report consists of 1138 pages; expect delays when accessing online documentation.

CRISPR

B





CpG mutations