



Optimizing care for patients with pustular psoriasis

Research Report and Summary from National Policy
Roundtable

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Table of Contents

Table of Contents	3
Executive Summary.....	4
Concluding remarks.....	6
1. Introduction	7
What is pustular psoriasis?.....	7
Disease occurrence and presentation.....	8
Impact on health	10
Health-related quality of life	10
Stigma.....	11
Morbidity, hospitalization and loss of life	12
Broader Impacts.....	13
Standards of care.....	13
Care pathways and guidelines	13
Unmet needs	13
Effectiveness of Current Treatments.....	14
Patient supports in Canada.....	15
2. Toward a better approach for Canadian PF-ILD patients	17
3. Supports identified through deliberation.....	19
4. Reflections from the 2022 Pustular Psoriasis Multistakeholder Policy Roundtable Discussions	22
Summary of discussion	22
Findings	23
Further Direction – What specific actions are needed and who will lead?.....	25
Concluding remarks	26
Appendix A – Approach to Policy Roundtable and Participants	27
Appendix B – Approach to Delphi Panel	29

Executive Summary

Pustular forms of psoriasis are chronic conditions set apart from other forms of psoriasis by the appearance of small (2-3 mm) bumps filled with pus (called pustules). As a chronic condition, it is associated with outbreaks (flareups) which can cause significant suffering and loss of function (e.g., walking, holding objects). It is a rare form of psoriasis, affecting ~1% of all patients with psoriasis (see Figure 1).

There are different ways to classify the disease, and the type of pustular psoriasis identified depends on where pustules form. As a rare condition, diagnosis relies heavily on the involvement of well-trained specialists who can set it apart from other skin conditions involving pustules. [Localized forms](#) are limited to small areas of the body, often palms and soles. A [generalized form](#) occurs over a much larger area of the body and may cause more illness-- patients with generalized pustular psoriasis can have life-threatening complications.

Of all forms of psoriasis (pustular and plaque), GPP causes the largest negative impact on health-related quality. Patients with pustular psoriasis, like other forms of psoriasis, are also stigmatized, and experience increases in hospitalization and a shortened life expectancy. It will also have a negative impact on worklife and productivity.

Because of the rarity of this condition, frontline healthcare practitioners may be less aware of its presentation and patients may receive inaccurate diagnoses and treatment. This highlights the need for appropriate levels of education and awareness; for GPP in particular, referral and appropriate communication between hospital and community-based practitioners may be a barrier to appropriate care and can have serious, even life-threatening consequences for patients.

Key Points

- Pustular psoriasis is a rare form of psoriasis; it is a chronic disease, with patients experiencing diminished quality of life, reduced productivity, stigma, and shortened life expectancy from disease.
- Optimal management of pustular psoriasis requires access to healthcare providers with highly specialized knowledge; the rarity of the condition means some patients may receive inappropriate diagnosis and treatment.
- Along with an increased knowledge of the disease and disease management by highly specialized practitioners, is an increasing recognition that others may be less informed, that standards of care are not widely accessible and that current patterns of care delivery and financing may be a disservice to patients and create worse experiences for patients and care providers.
- Key actions to optimize the future care of patients were identified through multistakeholder discussion. These include:
 - **Education and awareness for community-based dermatologists, and other frontline care providers.**
 - **The need to co-create Canadian clinical consensus guidance with patients and with a focus on the patient experience**
 - **The need to develop and finance networks to support excellent care – providing a platform for research, education and referral.**

Despite the considerable severity and burden of disease, further actions or strategies may be required to deal with gaps in care, and that aim to improve population health, reduce costs, and improve experiences for patients and clinicians.

To identify what supports are needed the most, the IHE conducted several multistakeholder (policy researchers, clinicians, clinical researchers, patients) evidence-informed discussions, including a survey of a larger group of patient representatives (n=3) and clinicians (n=15). Options were presented and discussed at an Institute of Health Economics, Virtual Multi-Stakeholder Policy Roundtable held on April 13, 2022 and with senior policymakers.

The Policy Roundtable discussion led to development of an action plan – to identify what supports are necessary for patients and potentially actionable by policymakers and larger communities of practice. The key findings are presented below:

Support required	What specific initiatives?	Who will lead this?
Education and awareness for community-based dermatologists (*and awareness for intensivists, internists, and hospitalists)	1) Webinars and remote education tailored to care providers: <ul style="list-style-type: none"> • direct-to-pharmacist, • direct-to-emergency physicians • direct-to-general practitioners • direct-to-dermatologists webinars. 2) Grand rounds (GPP) at local institutions	Canadian Dermatologists Association Thursday night Webinar Series Canadian Psoriasis Network hosts medical student roundtables The Canadian Skin Patient Alliance published first person narratives and can promote education through social media All stakeholders have a role in education at a local level
Creating Canadian clinical consensus guidance	Canadian consensus guidance developed with patient involvement and characterizing the patient journey	Dermatologist Association of Ontario (they have done a number of guidelines) Start with experts involved with previous guidance
Creating centres (networks) to optimize care	Local algorithms /pathways for how to access care need to be created, with patient involvement. A National vision for the role of networks and the essential role of tele-dermatology to connect healthcare providers requires development. ¹	Consensus based by region - needs to involve dermatologist leaders by province– requires consensus

¹ Note: CAPP, CSPA and CPN working on Teledermatology & Virtual Care project.

Concluding remarks

Further understanding and awareness of pustular psoriasis, coupled with its emerging evidence regarding the effectiveness of new approaches to treatment has created the need for innovative approaches to the care of patients with pustular psoriasis. The IHE Round Table discussion identified three priority actions to support patients: education initiatives for healthcare providers; consensus guidance; and the creation of specialized care networks to appropriately triage, treat and manage patients. The latter will require significant support from policymakers as it could challenge the current model of care delivery and financing.

1. Introduction

What is pustular psoriasis?

Pustular forms of psoriasis are chronic conditions set apart from other forms of psoriasis by the appearance of small (2-3 mm) bumps filled with pus (called pustules). Although not nearly as common as plaque psoriasis (see next section), patients experience similar pain and itch from this skin condition. There are different ways to classify the disease, and the type of pustular psoriasis identified depends on where pustules form. As a rare condition, diagnosis relies heavily on the involvement of well-trained specialists who can set it apart from other skin conditions involving pustules. [Localized forms](#) are limited to small areas of the body, often palms and soles. A [generalized form](#) occurs over a much larger area of the body and may cause more illness—patients with generalized pustular psoriasis can have life-threatening complications (Box 1). While it is unknown if causes of these two forms of pustular psoriasis are related, some genetic factors and environmental conditions have been identified, including smoking, stress, infections, and exposure to certain medications.²

Box 1 Generalized Pustular Psoriasis – a patient’s journey

Jessica is a 34 year-old woman from Northern Ontario who was once again experiencing a rash on her hands and arms that her doctor said was a kind of dermatitis. When she experienced this before, it was winter, and she could go out in the with a hat and mittens; now, she now found herself in the middle of summer, and feeling more exposed. Not only did she not want anyone to see her with these blemishes, her palms were starting to get sore and this was making it difficult to work in the garden and ride her bicycle.

Once again, she found herself at her doctor’s office. She was never sure what caused the rash but she knew this time it was not going away. She felt, in fact, it was beginning to worsen. While she had been using topical corticosteroids to try and get rid of the rash, her physician suggested she could get more rapid relief from taking an oral steroid. She happily took her new medicine and her condition improved. She was told by her physician that she would need to slowly reduce the dose of her oral steroid—this required her to take a slightly more complicated regimen of 2 fewer pills every three days until she was finished. She was told doing this would avoid unwanted side effects from stopping quickly.

Happy this was over, Jessica noticed she started to feel unwell. She had started to feel chilled, even having a mild fever. She started to experience pimples on her abdomen that were worsening and spreading all over her body; each one felt like a small fire. She knew something wasn’t right. She seemed to be breathing more quickly, and her heart rate was up and she lacked an appetite. Something was wrong. She managed to reach her doctor who told her to go to an emergency department.

When she arrived, the bumps had grown in size and number. She was red all over her trunk and the lesions had spread everywhere. They had spread to her legs. She was now in severe pain. The emergency room physician diagnosed an infection. She felt it was serious enough to admit Jessica to the hospital.

In the hospital she was given intravenous antibiotic therapy, and fluid replacement. Her fever and chills continued for 2 days and her rash only became worse. A series of tests were ordered and a dermatologist was consulted. With some additional laboratory tests, the dermatologist was able to confirm a diagnosis of *generalized pustular psoriasis*. She explained it is a disease characterized by flares—often once yearly or more – and the flares could be very serious. They could lead to a number of serious complications. Patients with serious flares have even been hospitalized.

Jessica was told her family doctor would need to refer her to a dermatologist for ongoing monitoring—at least every 3 months—. She would also need to initiate drugs which are often effective. If she didn’t respond well to these options, there are additional options, although some are very expensive and not always covered through insurance. She was told almost all of these drugs require contraception- and that it may be difficult to plan a pregnancy until she has had more experience with the condition.

² Adapted from: S. Kharawala et al., “The Clinical, Humanistic, and Economic Burden of Generalized Pustular Psoriasis: A Structured Review,” *Expert Rev Clin Immunol* 16, no. 3 (March 2020): 239–52, <https://doi.org/10.1080/1744666x.2019.1708193>; J. J. Crowley, D. M. Pariser, and P. S. Yamauchi, “A Brief Guide to Pustular Psoriasis for Primary Care Providers,” *Postgrad Med* 133, no. 3 (April 2021): 330–44, <https://doi.org/10.1080/00325481.2020.1831315>.

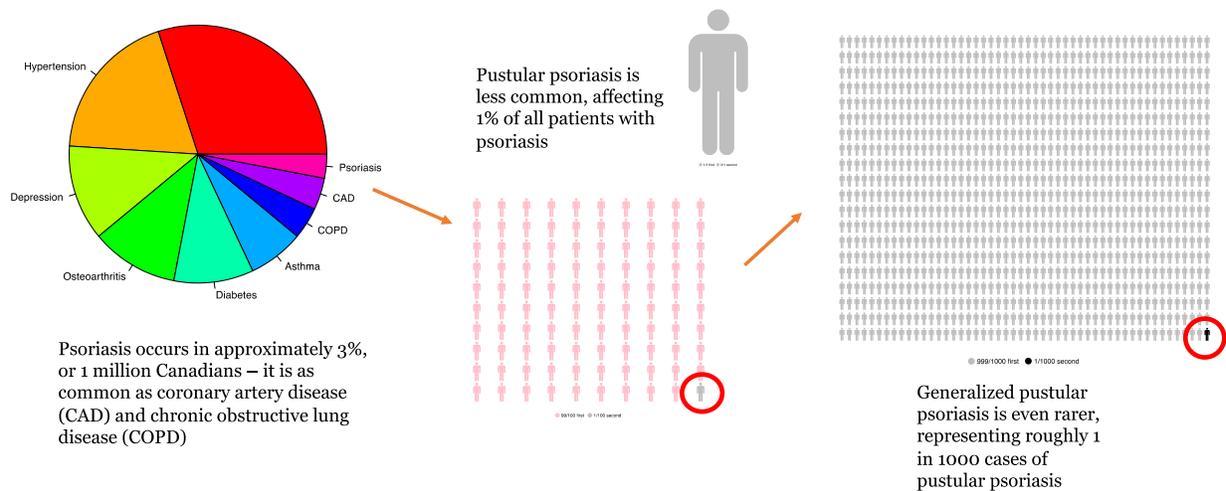
Disease occurrence and presentation

Pustular psoriasis occurs in two forms; the vast majority of individuals have a localized form where pustules erupt on the palms and feet (Figure 2, called palmoplantar pustulosis, PPP) or at the tips of fingers and toes (acropustulosis, or acrodermatitis continua of Hallopeau, ACH). As a chronic condition, it is associated with outbreaks (flareups) which can cause significant suffering and loss of function (e.g., walking, holding objects). It is a rare form of psoriasis, affecting ~1% of all patients with psoriasis (see Figure 1).

A second, more rare form of pustular psoriasis is a generalized pustular psoriasis (GPP) where pustular lesions rapidly extend to the trunk and limbs (Figure 2). It also results in flareups of disease with periods of partial or complete remission. Although it is estimated to be 1 in 1000 of all cases of pustular psoriasis, it is considerably more disabling. Of all forms of psoriasis (pustular and plaque), GPP causes the largest negative impact on health-related quality (Figure 3).

Patients experiencing their first onset of pustular psoriasis are typically working-age adults although it can occur in infants and children. Patients with GPP have often had a past diagnosis of plaque psoriasis (and can have both at the same time).³ Symptoms can resolve with limited intervention, or, especially in the case of GPP, may require urgent care. Complications in GPP patients such as infection and electrolyte imbalance have even led to hospitalization and death, in susceptible individuals.

Figure 1 Pustular psoriasis is a rare form of psoriasis



Adapted from ⁴; **Abbreviation:** COPD: chronic obstructive pulmonary disease; CAD: coronary artery disease.

³ R. Uppala et al., “Autoinflammatory Psoriasis²-Genetics and Biology of Pustular Psoriasis,” *Cell Mol Immunol* 18, no. 2 (February 2021): 307–17, <https://doi.org/10.1038/s41423-020-0519-3>.

⁴ “Chronic Disease Management in Primary Health Care: A Demonstration of EMR Data for Quality and Health System Monitoring,” n.d., 16; Crowley, Pariser, and Yamauchi, “A Brief Guide to Pustular Psoriasis for Primary Care Providers.”

Figure 2 Appearance of localized and generalized forms of pustular psoriasis

2a) Localized (palmoplantar) pustular psoriasis

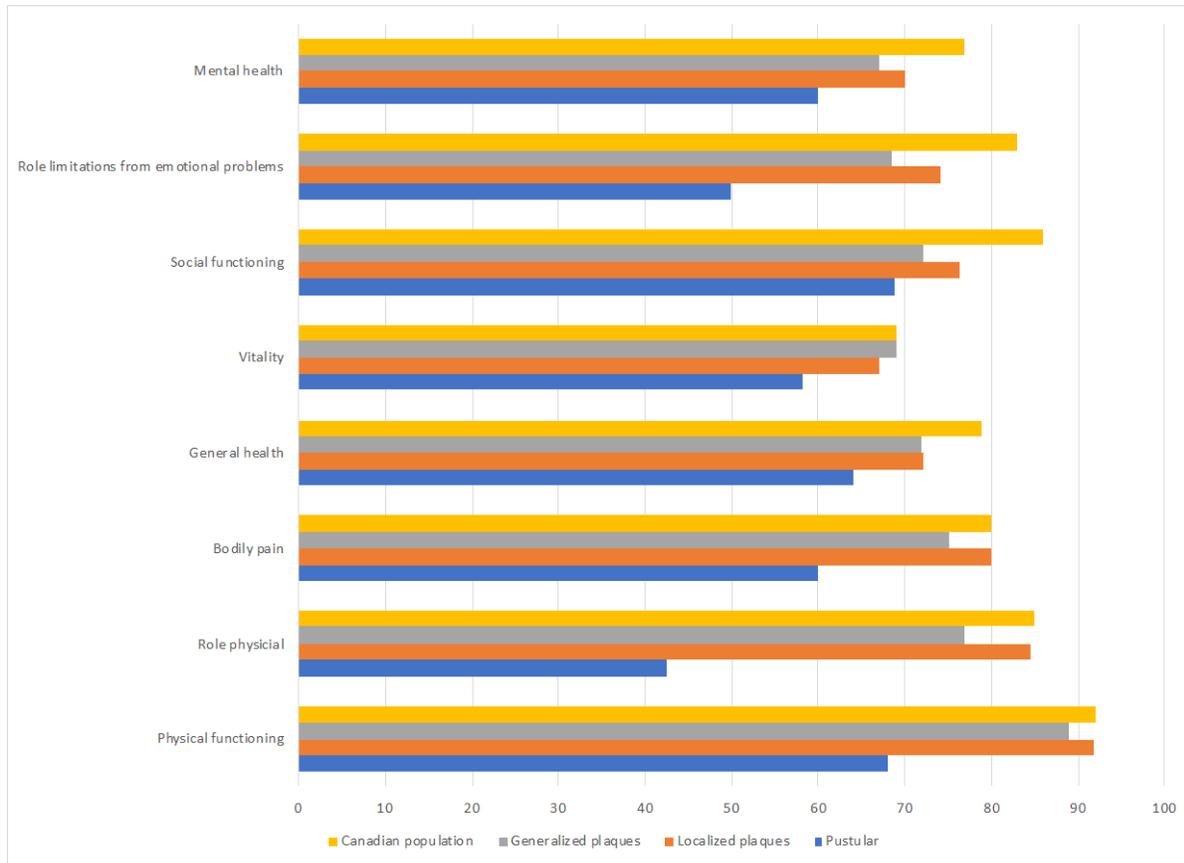


2b) Generalized pustular psoriasis



Images from
<https://dermnetnz.org/topics/palmoplantar-psoriasis-images>
<https://dermnetnz.org/topics/generalised-pustular-psoriasis-images>

Figure 3 Health-related quality of life associated with different types of psoriasis



Adapted from ⁵. **Notes:** Canadian scores are sex-standardized based on individuals 35-44 years-old.

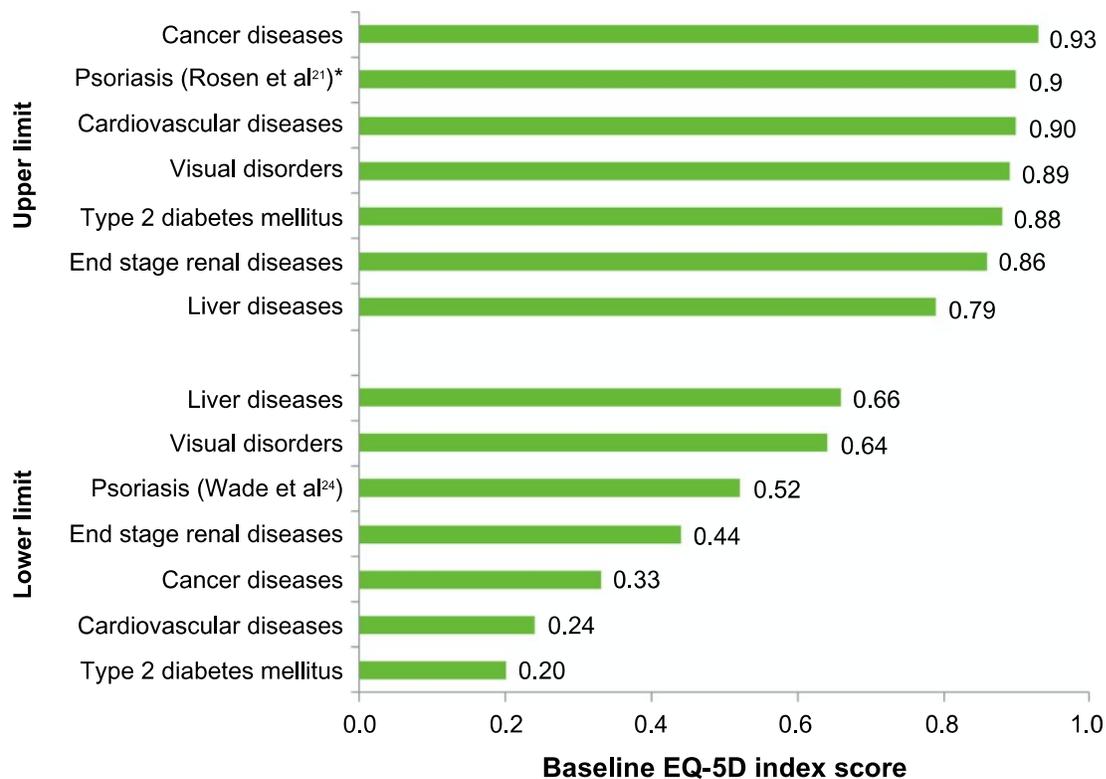
Impact on health

Health-related quality of life

The impact of all forms of psoriasis (i.e., pustular and plaque) on health-related quality of life is well documented. A comparison of the impact of psoriasis on key domains of health using a standardized tool showed psoriasis can be perceived as having a worse impact on quality of life than liver diseases (such as cirrhosis, hepatocellular carcinoma, and liver transplant) as well eye diseases (such as glaucoma, age-related macular degeneration, and cataracts). These key domains of health include patient mobility, ability to self-care, ability to engage in usual activities, level of pain and discomfort, and levels of anxiety or depression associated with illness. (Figure 3)

⁵ F. Sampogna et al., “Measuring Quality of Life of Patients with Different Clinical Types of Psoriasis Using the SF-36,” *British Journal of Dermatology* 154, no. 5 (2006): 844–49, <https://doi.org/10.1111/j.1365-2133.2005.07071.x>.

Figure 4 Impact of psoriasis on health-related quality of life compared to other chronic conditions



Adapted from⁶. **Notes:** scale: 0= dead; 1= full health. *Rosen et al reported the utility score to one decimal place (all other studies reported to two decimal places). Rosen and Wade were included in order to distinguish them from the other psoriasis articles, highlighting that these only reflect the findings from two of the papers. The study by Wade measured health-related quality of life in patients with moderate to severe psoriasis who required infliximab treatment. **Abbreviation:** EQ-5D: EuroQoL five dimensions patient reported outcome instrument.

These estimates also reflect the majority of psoriasis patients, who live with mild to moderate plaque psoriasis rather than those with the rarer, pustular forms, which can be even more disabling. Patients have consistently reported pustular psoriasis, when compared to any other forms of psoriasis, leads to a 25-30% decrease in perceived levels of physical functioning (e.g., ability to walk, bath, and climb stairs), vitality (perception of pep and energy level), and mental health (mood, anxiety and level of happiness, Figure 4).

Stigma

Despite these poor measures of health-related quality of life, they do not reflect the additional stigma and feelings of loss of control reported by patients with psoriasis. In a recent survey of Canadian patients to support a recent drug submission for plaque psoriasis⁷, approximately one-third of patients reported feelings

⁶ Nina Karaoghlanian et al., “A Systematic Literature Review to Compare Quality of Life in Psoriasis with Other Chronic Diseases Using EQ-5D-Derived Utility Values,” *Patient Related Outcome Measures*, July 2015, 167, <https://doi.org/10.2147/PROM.S81428>.

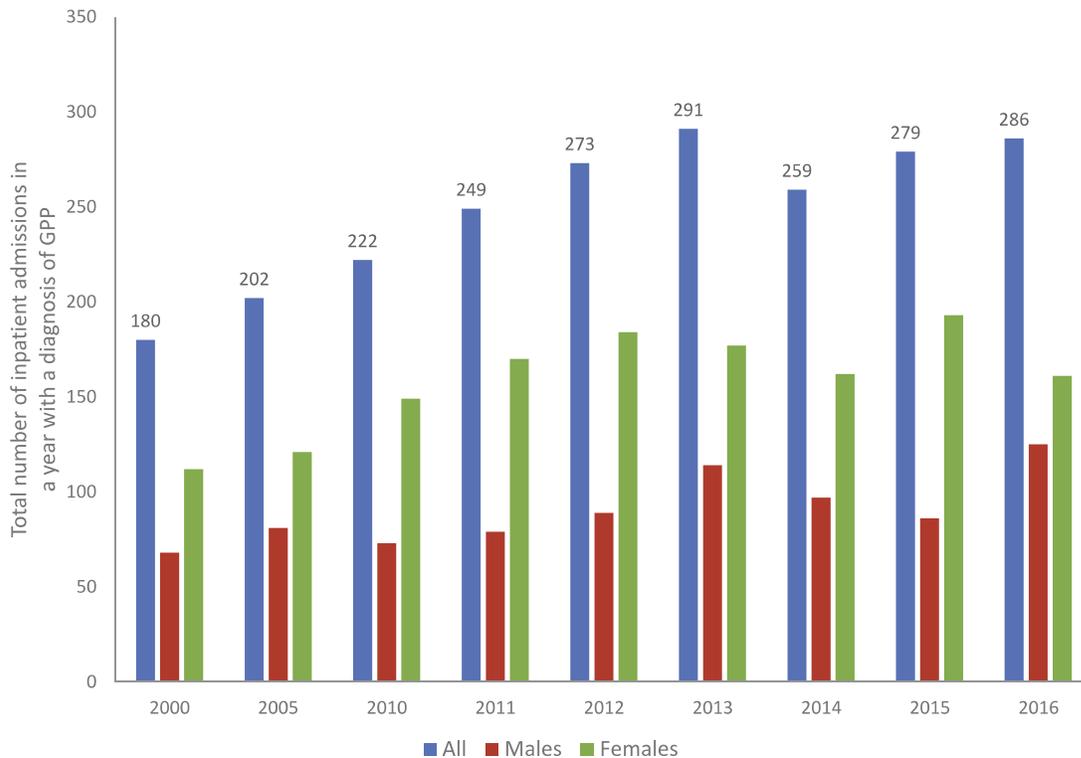
⁷ Canadian Agency for Drugs and Technologies in Health, “CADTH Common Drug Review Patient Input: CERTOLIZUMAB PEGOL (CIMZIA) for Moderate to Severe Plaque Psoriasis,” December 3, 2018, <https://www.cadth.ca/sites/default/files/cdr/relatedinfo/SR0587%20Cimzia%20-%20Group%20Patient%20Input%20->

of embarrassment, loss of sleep, problems with intimacy, and a negative effect on self-confidence associated with their psoriasis. Pustular psoriasis may carry additional stigma, particularly in patients who are (incorrectly) concerned with having infectious pustules and transmitting infection to others. They may similarly be stigmatized by those fearful of receiving an infection or concerned about the safety of others.

Morbidity, hospitalization and loss of life

Also not captured in measures of health-related quality of life are the various comorbid conditions that may also be prevalent, including pregnancy, higher rates of inflammatory bowel disease, cardiovascular disease, and arthritis. Severe forms of psoriasis are associated with an increased risk of mortality⁸, reducing the average life expectancy of male and female patients by 3.5 and 4.4 years, respectively. GPP is also associated with significant numbers of hospitalizations, and disproportionately affects women. Data from Germany suggests 250-300 patients are admitted to hospital every year with GPP (Figure 5).

Figure 5 Risk of hospitalization associated with severe forms of psoriasis (data from Germany)



Adapted from ⁹.

⁹ June 2028%2C%202019.pdf.

⁸ Joel M. Gelfand et al., “The Risk of Mortality in Patients with Psoriasis: Results from a Population-Based Study,” *Archives of Dermatology* 143, no. 12 (December 2007): 1493–99, <https://doi.org/10.1001/archderm.143.12.1493>.

⁹ Kharawala et al., “The Clinical, Humanistic, and Economic Burden of Generalized Pustular Psoriasis: A Structured Review.”

Broader Impacts

Like other forms of psoriasis, pustular psoriasis, which leads to reductions in vitality, mental energy and functionality, will also have a negative impact on patients who work.¹⁰ The impact is twofold: first, it may affect an individual's ability to perform physical tasks or stay focused; and secondly, it may make employees self-conscious, worried, or upset about their own ability to work and the perceptions of others.

Standards of care

Care pathways and guidelines

Although updated in 2016, current Canadian guidelines address treatment of palmoplantar pustular psoriasis (affecting hands and soles of feet) but not GPP. Treatment recommendations include the use of oral retinoids (e.g., oral acitretin) with or without phototherapy, calcipotriol topical therapy, and cyclosporine.¹¹ The guidance document also calls attention to the role of oral corticosteroid withdrawal and the use of TNF inhibitors as causes of pustular psoriasis. It recommends that patients who develop pustular psoriasis while receiving an TNF inhibitors (when it is to control another autoimmune disease), can continue use of the TNF inhibitors with topical therapies (calcipotriol, corticosteroids, or both), with or without phototherapy, being used to control disease.

More recent International Guidelines addressing GPP are from Japan (2018)¹² and the US (2012). Each guideline describes treatment approaches to pustular psoriasis in adults, pregnancy, and children based on a systematic review of available evidence. Tailoring treatment to the unique needs of patients, and considering side effect profiles and co-morbidities (such as use cetolizumab in pregnancy), or existing conditions (perimenopause / menopause) needs to be considered when making treatment decisions.¹³ Patients will often require combinations of therapies, which can increase the complexity and cost of treatment.

A recurring theme in clinical guidance documents are the very low levels of evidence available to inform decisions—typically non-experimental descriptive studies, such as comparative studies, correlation studies, or case-control studies (Figure 6). This means interventions may be given without strong proof-of-concept or approval from Health Canada (i.e., off-label treatments). Neither guideline provides guidance on what non-drug or other healthcare system supports might be required for patients.

Unmet needs

Like other chronic, autoimmune conditions, the needs of patients with pustular and other forms of psoriasis, are more complex. Patients must first have access to educational resources, a dermatologist, and potentially a larger multidisciplinary team so that they and their families can best understand short and long-term treatment goals. Ill-defined goals and the lack of an ability to achieve them may lead to patient dissatisfaction,

¹⁰ Related reference: www.workingitout.ca A report on the experiences of people with psoriatic disease in the Canadian workplace

¹¹ Canadian Psoriasis Guidelines Addendum Committee et al., “2016 Addendum to the Canadian Guidelines for the Management of Plaque Psoriasis 2009,” *Journal of Cutaneous Medicine and Surgery* 20, no. 5 (September 2016): 375–431, <https://doi.org/10.1177/1203475416655705>.

¹² Hideki Fujita et al., “Japanese Guidelines for the Management and Treatment of Generalized Pustular Psoriasis: The New Pathogenesis and Treatment of GPP,” *The Journal of Dermatology* 45, no. 11 (November 2018): 1235–70, <https://doi.org/10.1111/1346-8138.14523>.

¹³ “Canadian Psoriasis - CAPP - Baring It All,” accessed April 7, 2022, <https://www.canadianpsoriasis.ca/en/resources/reports/baring-it-all>.

suboptimal adherence, and ultimately, poorer quality of life.¹⁴

Treatment goals for patients typically consider the severity of the condition, access to drug and phototherapy services, comorbidities and potential side effects from treatment, along with individual patient preferences.¹⁵ In Canada, many of these factors have been identified as unmet needs.¹⁶ A Canadian Association of Psoriasis Patients access to care report recommended strategies to improve access to dermatologist care, phototherapy services and medications for patients with all forms of psoriasis. These were identified as critical for improving the lives of patients with psoriasis.¹⁷

Recommendations to address wait times for dermatology services have included the more widespread support for medical students, adoption of tele-dermatology services, specialized training of family physicians and nurse practitioners, and the adoption of rapid-access psoriasis clinics.¹⁸ Increased disease awareness, clinical education and practice guidelines, more equitable and increased access to available treatments, and the continued use of support programs for patients and families have also been identified as strategies to address the ongoing needs of psoriasis patients in Canada, and internationally.

Other potential unmet needs include education of community-based dermatologists; for GPP in particular, referral and appropriate communication between hospital and community-based practitioners which if not conducted properly may be a barrier to appropriate care.

Effectiveness of Current Treatments

A number of treatment options for plaque psoriasis have been used to treat pustular psoriasis and include oral retinoids (e.g., acitretin), disease modifying agents (e.g., cyclosporine and methotrexate), apremilast, phototherapy, and oral corticosteroids. An international survey of dermatologists also identified a number of biologics that have been used to treat generalized pustular psoriasis, including infliximab, adalimumab, etanercept, golimumab, certolizumab pegol, secukinumab, ixekizumab, brodalumab, and guselkumab.¹⁹

Despite the number of potential treatment options for GPP, robust evidence to support their use is largely

¹⁴ April W. Armstrong et al., “Undertreatment, Treatment Trends, and Treatment Dissatisfaction among Patients with Psoriasis and Psoriatic Arthritis in the United States: Findings from the National Psoriasis Foundation Surveys, 2003-2011,” *JAMA Dermatology* 149, no. 10 (October 2013): 1180–85, <https://doi.org/10.1001/jamadermatol.2013.5264>; “Pustular Psoriasis: Treatment Options,” accessed April 7, 2022, <https://www.aad.org/public/diseases/psoriasis/treatment/genitals/pustular>.

¹⁵ Lakshi M. Aldredge and Melodie S. Young, “Providing Guidance for Patients With Moderate-to-Severe Psoriasis Who Are Candidates for Biologic Therapy,” *Journal of the Dermatology Nurses’ Association* 8, no. 1 (January 2016): 14–26, <https://doi.org/10.1097/JDN.0000000000000185>.

¹⁶ The Economist Intelligence Unit, “Encouraging Policy Action to Address the Psoriasis Challenge” (The Economist Intelligence Unit Limited, 2017), <https://eiuperspectives.economist.com/sites/default/files/Eli%20Lilly%20%20-%20Encouraging%20policy%20action%20to%20address%20the%20psoriasis%20challenge.pdf>; Canadian Association of Psoriasis Patients, “Pso Serious 2018:A Report on Access to Care and Treatment for Psoriasis Patients in Canada,” October 29, 2018, <https://www.canadianpsoriasis.ca/images/CAPPPreportOctober27finalclean.pdf>.

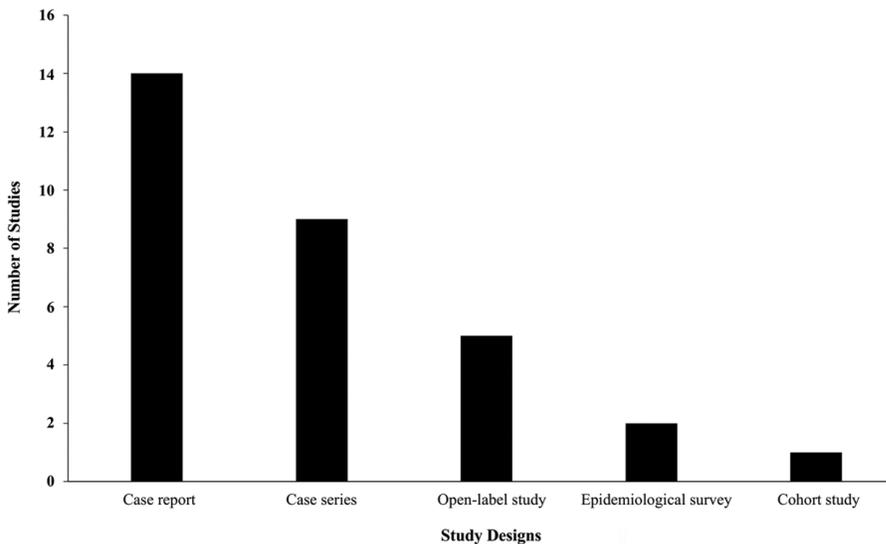
¹⁷ Canadian Association of Psoriasis Patients, “Pso Serious 2018:A Report on Access to Care and Treatment for Psoriasis Patients in Canada.”

¹⁸ The Economist Intelligence Unit, “Encouraging Policy Action to Address the Psoriasis Challenge”; Canadian Association of Psoriasis Patients, “Pso Serious 2018:A Report on Access to Care and Treatment for Psoriasis Patients in Canada.”

¹⁹ Bruce Strober et al., “Unmet Medical Needs in the Treatment and Management of Generalized Pustular Psoriasis Flares: Evidence from a Survey of Corrona Registry Dermatologists,” *Dermatology and Therapy* 11, no. 2 (April 2021): 529–41, <https://doi.org/10.1007/s13555-021-00493-0>.

lacking.²⁰ A 2018 systematic review of effective treatments in generalized pustular psoriasis identified 31 clinical studies, with no randomized controlled trials, and the vast majority case reports or case series (Figure 6).²¹ These types of studies do not allow for reliable comparisons between treatment strategies.

Figure 6 Study designs identified in a 2018 published systematic review of generalized pustular psoriasis treatments



Adapted from ²²

A review focused strictly on biologics for the treatment of pustular psoriasis (both localized and generalized forms), reported infliximab and ustekinumab have the most evidence of efficacy and safety and that recent evidence supports the use of IL-17 antagonists (secukinumab, brodalumab, and ixekizumab).²³ There is also some evidence to suggest patients are much less likely to discontinue biologics versus other treatment options.²⁴ The results of a German chart review indicated while 50% of patients are likely to discontinue any type of therapy at 14 months, this actually represented 36.0 months when biologics were employed compared to 6.0 months when non-biologics were used.²⁵ The introduction of new biologic treatment options has also been associated with improved quality of life for GPP patients in Japan, where the disease is most prevalent.²⁶ Despite this encouraging evidence, these treatments do not have a licensed indication in Canada for the treatment of any form of pustular psoriasis and are not funded through Canadian public insurance programs.

Patient supports in Canada

²⁰ L. L. Zhou et al., “Systemic Monotherapy Treatments for Generalized Pustular Psoriasis: A Systematic Review,” *J Cutan Med Surg* 22, no. 6 (November 2018): 591–601, <https://doi.org/10.1177/1203475418773358>; L. A. Falto-Aizpurua et al., “Biological Therapy for Pustular Psoriasis: A Systematic Review,” *Int J Dermatol* 59, no. 3 (March 2020): 284–96, <https://doi.org/10.1111/ijd.14671>.

²¹ Falto-Aizpurua et al., “Biological Therapy for Pustular Psoriasis: A Systematic Review.”

²² Zhou et al., “Systemic Monotherapy Treatments for Generalized Pustular Psoriasis: A Systematic Review.”

²³ Falto-Aizpurua et al., “Biological Therapy for Pustular Psoriasis: A Systematic Review.”

²⁴ C. Kromer et al., “Drug Survival in the Treatment of Generalized Pustular Psoriasis: A Retrospective Multicenter Study,” *Dermatol Ther* 34, no. 2 (March 2021): e14814, <https://doi.org/10.1111/dth.14814>.

²⁵ Kromer et al.

²⁶ K. Hayama et al., “Improved Quality of Life of Patients with Generalized Pustular Psoriasis in Japan: A Cross-Sectional Survey,” *J Dermatol* 48, no. 2 (February 2021): 203–6, <https://doi.org/10.1111/1346-8138.15657>.

Awareness and advocacy for patients with psoriasis in Canada is provided by the Canadian Skin Patient Alliance and the Canadian Association of Psoriasis Patients (www.canadianpsoriasis.ca). Additional support for patients is provided by the Canadian Psoriasis Network, regional programs, and individual patient support programs sponsored by pharmaceutical companies that manufacturer new agents for psoriasis. The mission of these organizations is described below (Box 1):

Box 2 Canadian organizations dedicated to patients with psoriasis (alphabetically)

- **Canadian Association of Psoriasis Patients**
Mission is to be a resource for psoriatic patients and their families to advocate for improved patient care and quality of life.
- **Canadian Psoriasis Network**
Mission is to enhance the quality of life of people living with psoriasis and psoriatic arthritis by providing current information on research and treatment options and by working with others to build awareness and advocacy about the complexities of these conditions (<https://www.canadianpsoriasisnetwork.com/about-us/>)
- **Unmasking Psoriasis**
Saskatchewan-based organization creating awareness about living with psoriasis (<http://unmaskingpsoriasis.org/>)
- **Psoriasis Newfoundland**
Newfoundland-based patient support and advocacy organization for people suffering from psoriasis and psoriatic arthritis in Newfoundland and Labrador.
(<http://psoriasisnl.com/about.html>)
- **Canadian Skin Patient Alliance**
Mission is to improve the health and well-being of people across Canada affected by skin, hair, and nail conditions through collaboration, advocacy, and education.

There are also numerous commercial patients support programs (see <https://www.canadianpsoriasis.ca/en/treatment/medication-access/patient-support-programs> for a complete list) that also support patients treated with currently licensed products for psoriasis.

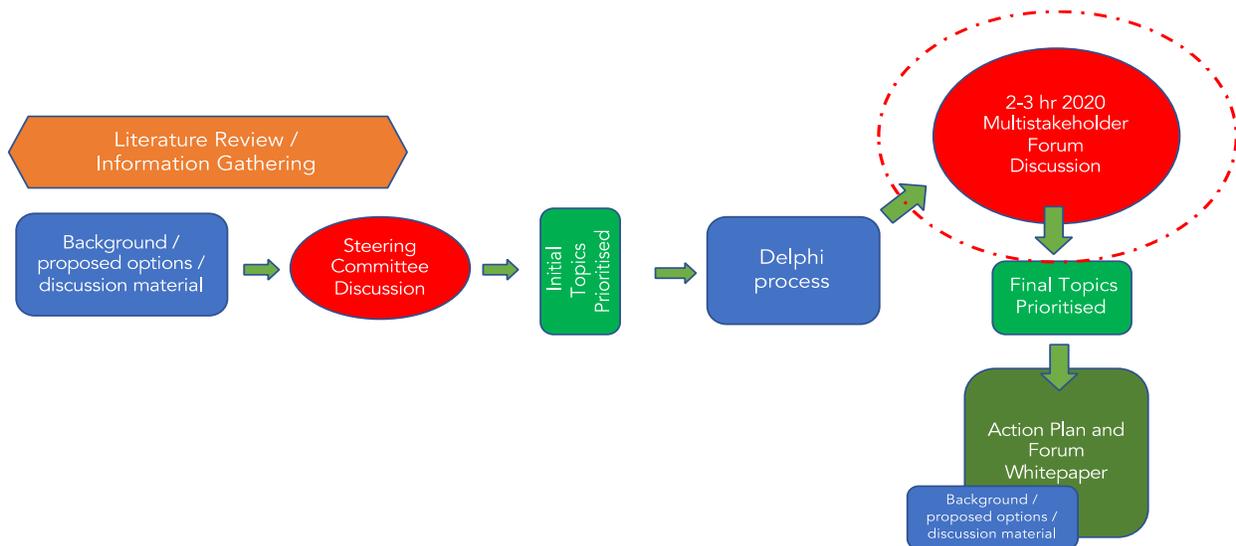
2. Toward a better approach for Canadian Pustular Psoriasis Patients

The Institute of Health Economics facilitated several ongoing discussions to identify key priorities and discuss research, practice and policy priorities in order to identify what further steps are needed to improve patient care and the patient journey for those with pustular psoriasis in Canada.

To begin with, the IHE held two multistakeholder (policy researchers, clinicians, clinical researchers, patients) evidence-informed steering committee discussions regarding potential priorities (“supports”) on November 30, 2021 and February 2, 2022. These supports could include research to address knowledge gaps, and strategic actions to aid clinical and payer policy, or any supports necessary for improving the capacity of health systems to optimize care of patients with pustular psoriasis. Several key priorities were identified in this meeting which were then further subject to a survey of a larger group of patient representatives (n=3) and clinicians (n=15). The survey used a modified Delphi²⁷ approach to elicit new priorities not identified and arrive at an initial ranking of priorities.

Figure 7 provides an overview of the process.

Figure 7 Approach to developing priority actions towards optimal patient care of patients with pustular psoriasis



The results were then presented at a second, multistakeholder Policy Roundtable held virtually on April 13, 2022 with the last session having an equal number of patients and clinicians. The objective of the invitation-only Roundtable discussion was to explore practical solutions and standards for providing better care to those with pustular psoriasis.

²⁷ Dmitry Khodyakov et al., “Practical Considerations in Using Online Modified-Delphi Approaches to Engage Patients and Other Stakeholders in Clinical Practice Guideline Development,” *The Patient - Patient-Centered Outcomes Research* 13, no. 1 (February 1, 2020): 11–21, <https://doi.org/10.1007/s40271-019-00389-4>.

The stated intent of the Round Table was to inform larger multi-stakeholder consensus on policy priorities ('supports') for patients with individuals with pustular psoriasis.

In particular, participants were asked to provide input on questions that would describe specific actions surrounding the priorities identified.

These questions included :

1. **What** education tools, research, changes to the organization of care, policies, or other key strategic actions **are needed to be implemented?**
2. **Who** would need to lead and/or contribute to achieve implementation of the strategic actions?
3. **What** special considerations for policy making and healthcare decision-making are needed?

The remainder of this report is organized as follows:

- **Section 2** provides an overview of the options identified by the steering committee and modified Delphi panel process
- **Section 3** provides a synopsis of the Multistakeholder Policy Roundtable reflecting on priorities from the modified Delphi process and some concluding remarks.
- **Appendix A** provides a description of Steering Committee, Workshop and Policy Roundtable attendees.
- **Appendix B** provides a description of the Delphi Panel approach

3. Supports identified through deliberation

As stated in Section 2, the overarching goal of engaging multiple stakeholders was to identify and prioritize ‘supports’ that are necessary for improving the capacity of health systems “to optimize care” of patients with pustular psoriasis. As a less well understood condition that is not frequently diagnosed, there may be opportunities to improve health care system readiness as newer treatments become available.

‘Supports’ is a broad concept and was defined for participants as “actions or strategies to deal with existing care gaps.”

Examples provided to participants included:

- Educational tools or guidance for patients, physicians, or policymakers
- Research to address important gaps in knowledge
- Changes to how care is organized, how patients are seen, or how care decisions are made
- Creating communities of practice or information technology tools to connect those involved with care (e.g., patients and physicians)

Similarly, “optimizing care” is a subjective concept. A definition of ‘optimizing care’ provided to participants was: “creating conditions for care that align with the triple aim of healthcare”. This includes:

- Improving health outcomes for those with pustular psoriasis
- Improving patient and care provider experiences from diagnosis to resolution of illness
- Reducing costs or keeping expenditures at a minimum

Beyond the “quadruple aim” in healthcare, there may be other considerations related to optimizing care that could include improving health care and informal care provider experiences, improving equity of access to care, improving social determinant of health, and improving healthcare system readiness.

A number of supports were identified by the Steering Committee. Two additional options were also offered by Panel members and included “creating networks to bridge providers” as well as “creating awareness for care providers of all available treatment options”.

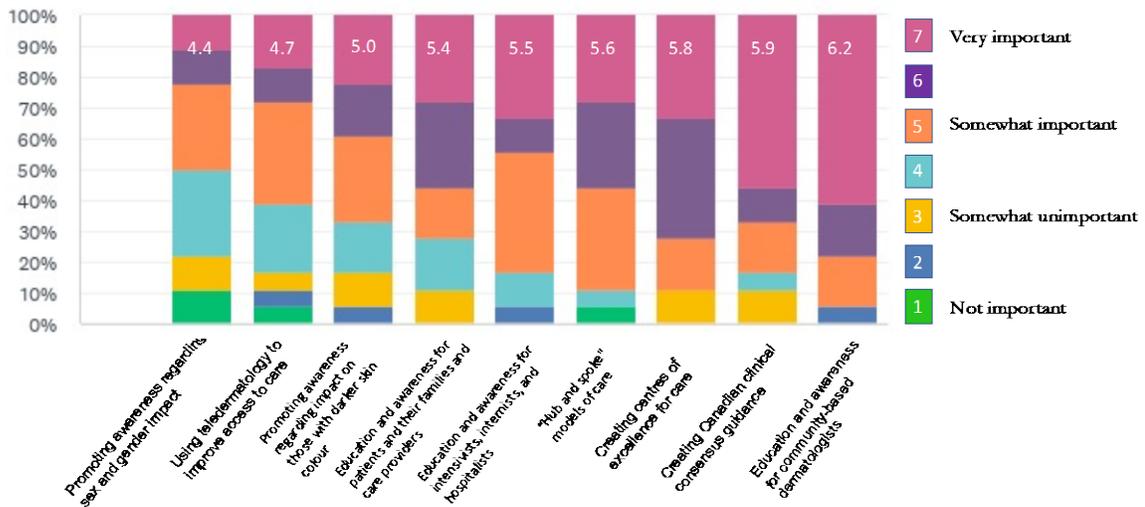
A description of these supports, and their accompanying rationale appears in Table 1 , below.

Table 1 Necessary supports identified through the Steering Committee and Multistakeholder Panel

Support (alphabetically)	Description	Rationale
Creating Canadian clinical consensus guidance for how to recognize and treat pustular psoriasis	Typically, peer-reviewed published guidance that is then disseminated through educational channels for physicians	Better knowledge about this condition will improve patient experiences, care provider experiences, and patient outcomes
Creating centres of excellence for care	Regional hubs that can work as referral centres but also as centres of research and support for patients	Centres of excellence can serve as regional hubs but can also aid with research activities to better understand disease, and patient experience, and optimize care experiences.
Education and awareness for community-based dermatologists	Education through courses, workshops, lectures, grand rounds and other established educational approaches for physicians	Better knowledge about this rare condition will improve care and decisions about referring to more specialized centres and the patient and care provider experience.
Education and awareness for intensivists, internists, and hospitalists		Better knowledge about this rare condition will improve patient experiences and outcomes
Education and awareness for patients and their families and caregivers	Education through ad campaigns, websites, and other established educational approaches for families and caregivers	Better knowledge about this rare condition will improve patient navigation, knowledge about available supports, and the patient experience.
Implementing "hub and spoke" models of care	Care providers in communities are given information on where to refer patients with the condition	Quick referral to a centre with more direct care experience will improve patient experience, outcomes and could reduce costs.
Promoting awareness regarding impact on a) gender and b) those with darker skin colour	A combination of educational modalities for clinicians, patients, families and caregivers	Awareness of impact on disadvantaged groups will improve patient and caregiver experiences.
Using tele-dermatology to improve access to care	Allowing consultation with other care providers through virtual care and teledermatology.	Tele-dermatology consultation can speed up diagnosis and appropriate referral and care and may reduce costs.

The results of the rating exercise for these items are presented in Figure 8. The three highest rated Items, based on level of importance, were: 1) Education and awareness for community-based dermatologists; 2) Creating Canadian clinical consensus guidance for how to recognize and treat pustular psoriasis; and 3) Creating centres of excellence for care. These Items were the focus of the final Multistakeholder Forum Discussion, and are discussed further in Section 4 (Page 22)

Figure 8 Relative importance of identified supports to optimize care in pustular psoriasis, via the modified Delphi procedure.



Aid to interpreting this figure:

- Participants, including patient representatives (n=3) and clinicians (n=15) were asked to provide a rating for each Item.
- The order of support options was randomized for each participant so that items did not receive a preferential rating due to list order.
- A weighted average for each Item was calculated by assigning a value from 1-7 for each Item, adding up the total value from all respondents, and dividing by the number of responses received for that Item. (i.e., a weighted average score)

4. Reflections from the 2022 Pustular Psoriasis Multistakeholder Policy Roundtable Discussions

The background and options described in Sections 1 and 3 were presented and discussed at an Institute of Health Economics, Virtual Multi-Stakeholder Policy Roundtable held on April 13, 2022. Round Table Participants (n=8, See Appendix A – Approach to Policy Roundtable and Participants) as well as at individual meetings with senior policymakers held after the Roundtable (n=2).

The Roundtable followed a format of reviewing key messages from the background document (Section 1), the approach taken to deliberation, followed by a presentation of the rating of supports in Section 3. Roundtable attendees participated in a facilitated discussion to determine what supports were of highest priority. This discussion was then synthesized to facilitate further discussion and achieve broad consensus. The session was designed to support sharing and exchange of perspectives and information.

Summary of discussion

When discussing the supports and their rating, Round Table participants made the following general observations:

- The two additional supports nominated during the Delphi procedure (“Create networks” and “Create awareness of treatment options”) were largely felt to be already addressed in other items presented (i.e., “Implement hub and spoke models” and “Create Canadian clinical consensus guidance”, respectively).
- The relative weighting of the supports may be reflective of the larger proportion of clinicians who were involved in both the steering committee and the Delphi Panel.
- Supports to address issues related to capacity and wait times may have been underweighted, as regions with little capacity were also underrepresented throughout deliberation. This includes supports such as tele-dermatology and education for general practitioners, which may become more relevant in regions such as Saskatchewan, which have few (<10 for the province) dermatologists compared to regions such as Southern Ontario.

Findings

Policy Roundtable participants deliberated on key dimensions related to delivering optimal care for patients with pustular psoriasis in Canada including what key directions are needed and who is best suited to move them forward.

1. Education and awareness for community-based dermatologists

- Although education for community-based dermatologists was a recognized need, front-end providers may differ according to the capacity in any given area. As such, this education may need to be tailored to the needs of individual jurisdictions who may have fewer available community-based dermatologists, and where general practitioners or other front-end care providers (e.g., pharmacists) makes more sense.
- Similarly, education for health care professionals must be tailored to the patient experience and their journey through the healthcare system. This means education about generalized pustular psoriasis will become more relevant to emergency physicians, intensivists, and hospitalists whereas education about localized pustular psoriasis is more relevant for community-based practitioners. **In this sense, education and awareness for intensivists, internists, and hospitalists should be bundled into this item.**
- Educational initiatives will also need to be tailored to the type of healthcare professional which may be interested in different aspects of these diseases.
- It is recognized all stakeholders have a role in education, however all stakeholders will also benefit from having a detailed understanding of the patient's experience with this condition
- Education about pustular psoriasis is also part of a broader conversation about rare disease.

2. Creating Canadian clinical consensus guidance for how to recognize and treat pustular psoriasis

- In Canada, consensus guidelines in dermatology tend to be produced by a core group of individuals who have an academic interest in creating evidence-based standards.
- There is a recognized need for consensus guidance in this area, given a current lack of comprehensive guidance and a number of emerging studies that contain insights about diagnosis, the genetic basis of disease and highlighting the effectiveness of new treatments in this area.
- In addition to using a rigorous approach to the identification of evidence and values to support recommendations in clinical guidance, any new guidance will need to involve patients and fully characterize the patient journey, in keeping with new international standards about involving patients and the public in clinical guideline development.²⁸
- Given new standards in the approach to guideline development, the time and resources required to develop guidance should not be underestimated.

²⁸ “New Digital GIN Public Toolkit: Patient and Public Involvement in Guidelines.,” *GIN* (blog), September 30, 2021, <https://g-i-n.net/new-digital-gin-public-toolkit-patient-and-public-involvement-in-guidelines/>.

3. Creating centres of excellence for care

- As with education, the remit of a centre of excellence may vary according to jurisdictional capacity to deliver care; there was general consensus that most medical dermatologists are able to manage localized pustular psoriasis but that this may still be the focus of a specialized centre for those jurisdictions where medical dermatologists are more difficult to access.
- In order to be effective, centres must be able to accept patients from community dermatologists
- As all centres will be a hub for the management of generalized pustular psoriasis, they will also need to be linked to highly specialized hospitals where these patients must receive care, such as academic hospitals.
- Centres can play a central role in a hub and spoke model of care; in order to be useful, a clear referral pathway (i.e., a consensus ‘algorithm’) needs to be mapped according to the needs of any jurisdiction and special supports (e.g., remuneration) may be required in some jurisdictions to make these care and referral pathways feasible.
 - In this sense, it is important to establish that a province may not have a physical “centre” but rather a locus of expertise that is highly coordinated – **a network of care rather than a centre.**
 - In some jurisdictions, the use of a centres of excellence may more heavily rely on the use of tele-dermatology; this may also require support by policymakers for the care of patients through extra-provincial arrangements.
 - In this regard, some provinces still lack billing codes or policies surrounding tele-dermatology, making a centre-of-excellence model of care infeasible
- An overarching (National) vision will be an important starting point to build an infrastructure that is fit-for-purpose.
- Any centre-of-excellence model must consider the role health care providers outside of dermatology; this includes mental health and allied health professionals
- Similarly, there may be perverse incentives for care that require consideration in any centre-of-care model.

Further Direction – What specific actions are needed and who will lead?

Patient representatives observed that supports are often developed without the meaningful involvement of patients; and, therefore, the development of any support to improve patient lives should also involve patients.

Participants were then tasked to develop a list of concrete actions including who could lead them and how these might be accomplished. A preliminary list is presented in Table 2. It was recognized that there may be further options and that this list could be further refined or added to.

Table 2 Final recommendations for priority supports identified at the Multistakeholder Forum

Support required	What specific initiatives?	Who will lead this?
Education and awareness for community-based dermatologists (*and awareness for intensivists, internists, and hospitalists)	1) Webinars and remote education tailored to care providers: <ul style="list-style-type: none"> • direct-to-pharmacist, • direct-to-emergency physicians • direct-to-general practitioners • direct-to-dermatologists webinars. 2) Grand rounds (GPP) at local institutions	Canadian Dermatologists Association Thursday night Webinar Series Canadian Psoriasis Network hosts medical student roundtables The Canadian Skin Patient Alliance published first person narratives and can promote education through social media All stakeholders have a role in education at a local level
Creating Canadian clinical consensus guidance	Canadian consensus guidance developed with patient involvement and characterizing the patient journey	Dermatologist Association of Ontario (they have done a number of guidelines) Start with experts involved with previous guidance
Creating centres (*networks) to optimize care	Local algorithms /pathways for how to access care need to be created. A National vision for the role of networks and the essential role of tele-dermatology to connect healthcare providers requires development	Consensus based by region - needs to involve dermatologist leaders by province– requires consensus

Concluding remarks

Further understanding and awareness of pustular psoriasis, coupled with its emerging evidence regarding the effectiveness of new approaches to treatment has created the need for more awareness, as well as consistent and innovative approaches to the care of patients with pustular psoriasis. The IHE Multistakeholder Round Table Forum identified three priority actions to support patients: education initiatives for healthcare providers; consensus guidance; and the creation of specialized care networks to appropriately triage patients. The latter (networks) will require significant support from policymakers as it could challenge current model of care delivery and financing. For each of these initiatives, some initial feasible actions have been identified that are ultimately intended to support provincial jurisdictions in their delivery of optimal care to these patients.

Appendix A – Approach to Policy Roundtable and Participants

The Roundtable was an invitation-only event hosted by the Institute of Health Economics with support provided by Boehringer Ingelheim (Canada) Ltd./Ltée.

The event, invited participants, and Agenda are shown below:

How can we optimize the care of patients with pustular psoriasis?

April 13th – 2022 – 6:30pm – 9:00 EDT

The Institute for Health Economics (IHE) is conducting an invitational, virtual policy roundtable on pustular psoriasis on April 13th, 2022. This policy roundtable will seek to gain multi-stakeholder consensus on policy priorities and supports for patients with individuals with pustular psoriasis.

Key areas for discuss are in the following three areas:

- Education and awareness of community, based dermatologists, intensivists, internists and hospitalists
- Creating centres of excellence for care, hub and spoke models, and the use of tele-dermatology
- Creating Canadian clinical consensus guidance for how to recognize and treat pustular psoriasis

The purpose of this discussion will be to bring together multiple stakeholders from across Canada to review the current state of evidence and information regarding pustular psoriasis, examine provincial and regional policies related to this, and identify and prioritize necessary supports that are necessary for improving the capacity of health systems to optimize care of patients with pustular psoriasis.

In particular, participants will provide input into questions such as:

- What education tools, research, changes to the organization of care, policies, or other key strategic actions are needed to be implemented?
- Who would need to lead and/or contribute to achieve implementation of the strategic actions? and provide recommendations regarding special considerations for policy making and healthcare decision-making.

This invitational forum is being organized by the Institute of Health Economics (IHE) and is supported by and developed in partnership with Boehringer Ingelheim (Canada) Ltd./Ltée.

Approach: A workshop report will be conducted using the Chatham House rule (i.e. non attribution of feedback) . It is not expected that this will be a Consensus Report but a summary of discussion where areas of general agreement are noted along with areas where there are differing points of view.

Attendees

<p>Patient representatives Antonella Scali, Executive Director at Canadian Psoriasis Network Christian Boisvert-Huneault, Montreal, QC Margaret Peters, Director, Unmasking Psoriasis Rachael Manion, Executive Director at Canadian Skin Patient Alliance & Canadian Association of Psoriasis Patients</p>	<p>Healthcare providers Charles Lynde, Markham, ON Jaggi Rao, Edmonton, AB Jennifer Beecker, Ottawa, ON Ron Vender, Hamilton, ON</p>
<p>Institute of Health Economics Don Husereau, Ottawa, ON</p>	<p>Boehringer Ingelheim Roger Chai, Toronto, ON</p>

AGENDA

Major Objective:

To receive feedback and prioritize proposed supports for improving the care of patients with pustular psoriasis.

Agenda: *(meeting will be continuous – participants can take individual breaks as required).*

18:30 – 18:45

- Introductions – IHE and BI – Overview of Initiative
- Presentation by IHE on Key Findings in Pustular Psoriasis
- Review of Meeting logistics/format.

18:45 – 19:45

Major discussion areas

- a. Review of priorities (IHE presentation):
- b. Input on Key Recommendations related to supports identified :
 - *What?* – Elaborate on specific actions
 - *Who?* - Identify Necessary Leaders/Supporters/Coalitions
 - *Role for patient education/advocacy*

19:45 – 20:00 (break) – Group leaders to summarize key findings.

20:00 – 21:00 – Plenary

- Perspectives – comments on priorities and actions from patients, health care providers and policymakers
- Online polling and utilization of comment function and roundtable feedback to solicit additions/comments on recommendations.
- Prioritize/Probe areas of agreement/disagreement
- Closing remarks and Next Steps

For further information contact: Don Husereau, Senior Associate,
donh@donhusereau.com

Appendix B – Approach to Delphi Panel

OBJECTIVE

The overarching objective is to produce a comprehensive and prioritized list of supports that need to be addressed to optimize (i.e., meet the quadruple aim of reducing costs, improving patient and provider experiences, and population health) of individuals with pustular (generalized and PP) pustular psoriasis.

METHODS AND TIMELINES

Participants

The Delphi procedure will aim to capture the following perspectives

- Patients
- Patient advocates
- Dermatologic nurses
- Hospital administrators/HTA
 - Formulary committee director (hospital P&T)
- Chronic disease/mental health specialists
 - Experts that may consider the impact of underserved communities in skin care (sex, gender, skin colour)
- High-level Policymakers
- Academic derm. specialist
- Derms. that work in hospitals
- Regional dermatologist perspectives
- Allied health professionals (pharmacy?)

Process

A modified Delphi process (2) will be using SurveyMonkey and administered by DH

Round 1

Participants will be provided brief background information and shown a preliminary list of care gaps identified by the steering committee and asked to

- 1) comment on their perceived importance,
- 2) rank the importance of the item (on a Likert scale – 1-7, see Figure below as an example)
 - a. provide a rating of confidence in the ability to rate the item.
- 3) nominate additional issues/gaps.

Round 2[optional, depending on Round 1]

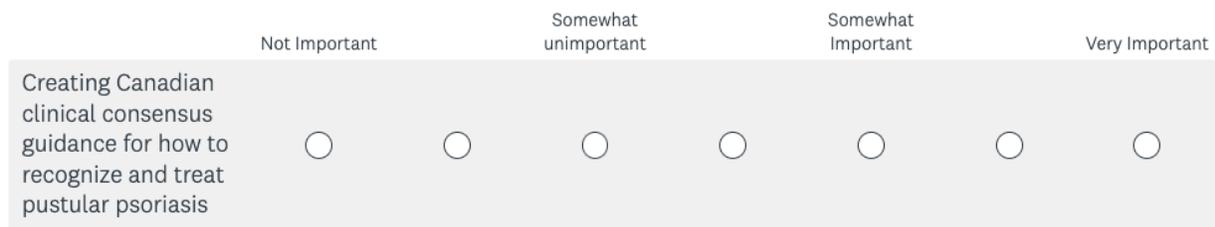
The items will then be synthesized and vetted by Steering Committee members before a second round of survey items is circulated. Participants in the 2nd round will again be asked to

- 1) comment on their perceived importance,
- 2) rank the importance of the item (on a Likert scale – 1-7)

The survey should take 10 minutes to complete in each instance. Support items will be randomized for each participant. A weighted average score for each support item will be calculated by adding the weights of the Likert values and dividing by the number of participant responders.

Figure 9 Example of Likert Scale used in the modified Delphi procedure.

* Rate the relative importance of each of the items



DRAFT EMAIL

Hi XXX,

I am currently participating in a project attempting to identify care gaps and potential solutions in individuals diagnosed with (generalized and palmoplantar) pustular psoriasis.

We are interested in capturing the opinions of others who may have important perspectives to share. The goal is to develop a comprehensive list of issues and solutions that may help to affect policy change.

As such, we were hoping to seek your input.

The Delphi survey will be administered by email and begin [date] and could require up to 10 minutes of your time in the first round. It is anticipated the survey will be repeated once 1-2 weeks after this time.

It is anticipated a published report will be made available in the spring, and if you wish, you could be acknowledged as a survey participant.

If you are interested in participating in the survey, please let me know by [DATE], and I will pass on your name to Don Husereau, who will send you an official invitation shortly.

Cheers,

XXX