Peristomal pyoderma gangrenosum (PPG) is a subtype of pyoderma gangrenosum (PG) that presents unique diagnostic and therapeutic challenges and opportunities. In this session, we will explore the question of whether PG at special sites warrants similar or distinct considerations in diagnostic evaluation and management.

In this session, we will cover 3 learning objectives:
1. Discuss best practices for the diagnostic evaluation of PPG
2. Describe special considerations in the approach and therapeutic ladder for PPG versus classic PG
3. Select best wound care strategies for PPG

The diagnostic evaluation of a patient with suspected PPG includes:
- PG diagnostic criteria (Su W et al, 2004, IJD, 43:790-800)
- Typical features on morphology:
  - violaceous, undermined border, cribiform ulceration -> characteristic scarring pattern.
  - often begin as erythematous to purpuric papule or pustule especially at mucocutaneous junction with stoma
  - almost all PPG occur wholly or partly behind the ostomy faceplate adhesive
  - 2/3 lie within millimeters or involve the mucocutaneous junction with stoma
- R versus G-type PG: Some investigators have subclassified PG into R (rapidly-expanding) versus G-type (indolent) subtypes, and this has been considered in cases of PPG. It is interesting to note that in the few studies that exist, R type PPG are associated with ulcerative colitis (UC) and involve both peristomal and median surgical wounds. G type PPG are associated with Crohn disease and typically have stomal involvement only.
- D/dx: - infection - cutaneous IBD or enterocutaneous fistula - contact dermatitis - vasculitis - ischemia (due to convex-type ostomy appliances which cause pressure necrosis/ pressure ulcer) - trauma (from ill-fitting ostomy appliance)
- To biopsy or not biopsy? Recommend to always biopsy as the differential diagnosis is broad and many studies have documented high misdiagnosis rate of PG (Weening et al, 2002 NEJM 347:1412-1418)

Demographics of PPG (summary of systematic review, 355 cases, 81 papers available on PubMed, MedLine, EMBASE)
- Mean age 47.5 years, F:M [2:1]
- Incidence with ostomy placement 0.6-1.5% in all ostomy series, higher rates (2-4.3% in ostomy series of IBD patients only)
- Risk factors: younger age, wound infection (both cutaneous and intra-abdominal), bowel obstruction, hernia, female, increased BMI, concurrent autoimmune disease
- Onset of PPG after ostomy: mean 23 months (range 2 days – 26 years)
- Stoma types associated with PPG: ileostomy (81%), colostomy (14%), urostomy (3%)
- Associated disease: Crohn disease 50% (40% concurrence rate of PPG + Crohn disease flare)
  Ulcerative colitis 31% (20% concurrence rate of PPG + UC disease flare)
  Other 19% (38% concurrence rate of PPG + flare of condition)
- High recurrence rate: up to 1/3 of patients in most series

Pathogenesis of PG: there is no available data to suggest that the pathogenesis of PPG is distinct from PG. Aberrations in neutrophil recruitment & homeostasis, mediated by neutrophils, T cells, genetics, cytokines, are the primary pathways leading to any neutrophilic dermatosis.

Best practices for PG management:
- Important concept -> treatment of PG must include BOTH wound management + treating inflammation (Bennett, 2000, Medicine, 79:37-46)
- Additional goals include reducing pain, evaluation/ treatment for associated systemic condition
- It is significant to note that treatment for associated systemic condition may not be sufficient to heal the PPG.
Ongoing need to establish evidence and best practices through collaborative study of rare skin diseases.

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<th>Key points of the talk:</th>
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<tr>
<td>PG at special sites: unique diagnostic and treatment considerations, opportunities for creative and evidence-based management.</td>
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<td>Management of PPG: wound care + treating/ reducing inflammation are equally important aspects. Both must be addressed.</td>
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<td>Individualized wound care and collaboration with ostomy nursing expert and colorectal surgeon is critical to successful wound healing.</td>
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<td>Ongoing need to establish evidence and best practices through collaborative study of rare skin diseases.</td>
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**PPG treatment: addressing inflammation**
- Summary of systematic review (355 cases, 81 papers)
- Topicals: first line is corticosteroids (caution: steroid atrophy from occlusion), alternative calcineurin inhibitors, IL corticosteroids
- Topical STOP-GAP trial looked at clobetasol versus tacrolimus (multiple strengths) and showed that both are effective as monotherapy for PG with slow healing times. Smaller ulcer size at the start of treatment was associated with better outcomes.
- Systemics: first line therapy is corticosteroids with an important alternative being cyclosporine. The recent Systemic STOP-GAP trial looked at prednisolone (0.75 mg/kg/day) versus CSA (4mg/kg/day) for both 6 week and 6 month healing and found that they were comparable, with slight trend for faster healing with CSA. Importantly, both treatment groups had significantly high rate of adverse events (2/3 of patients) with higher infection rates in patients taking systemic corticosteroids.
- Additional agents: anti-neutrophilic agents (dapsone) and biologics (primarily anti-TNF alpha inhibitors) have shown some benefit for PPG, limited case reports only
- Systemic review showed that combination therapy (systemic + topical used together) leads to faster healing times.
- Systemic only therapy may be needed depending on the ostomy care regimen
- Treatment may be needed for 3-56 weeks, mean 7-12 weeks

**PPG management: wound care**
- Summary of systematic review (355 cases, 81 papers)
- Special considerations for wound care management in PPG
  - maceration, fecal/ urine exposure
  - contact dermatitis: both irritant and allergic. A significant majority of patients with ostomy have irritant dermatitis.
    - Always consider a concomitant irritant dermatitis in conjunction with PPG as healing of the PPG may depend on treatment of the concurrent irritant dermatitis.
    - Consider patch testing: cleaning or prep agents (fragrance, cleanser, preservatives), glues, adhesives, medication (vehicle – propylene glycol), steroids, foam/ plastics. Consider cutting ostomy appliance into parts and using these in USE or traditional patch testing analysis.
  - increased risk of steroid atrophy (due to occlusion under ostomy appliance)
  - unique vehicles (spray – i.e fluticasone nasal spray or beclometasone inhaler) may enable use of topical steroids under adhesives and also special wound dressing types may be employed even under ostomy
    - i.e. fluticasone nasal spray, beclometasone respiratory inhaler
    - foam (i.e. Allevyn), hydrocolloid (i.e. Duoderm), calcium alginate, Hydrofera blue (gentian violet)
  - powders: sucralfate (absors maceration), dapsone (crushed tablets)
  - consider soaks such as Domeboro’s or Dakin for disinfection and maceration (avoid direct soaking of stoma)
  - ostomy nurse collaboration: to determine alternative sizing (many patients with ostomy change sizes over time)

**Surgical considerations:**
- Split-thickness skin graft may be helpful. There is a notable case report of a mesh assisted STSG used in a patient with intractable pain due to an almost circumferential PPG – this patient was terminally ill which was a major factor in placing mesh under the graft.
- Ostomy closure: 15 cases described in the literature -> 100% healing
- Stoma revision/ relocation: 36 cases described in the literature -> 86% healing but very high rate of creating new PPG at the new location of the stoma.
- Debridement: 34 cases described in the literature -> 53% healing