原文題目(出處)：	Medical management update: Peutz Jeghers syndrome
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內文：

前言

1. Peutz Jeghers syndrome (PJS) is an autosomal dominant disease characterized by polyps and mucocutaneous pigmentation that typically manifests in childhood and early adult hood, with a common presentation of bowel obstruction and severe abdominal pain.
2. Diagnostic criteria include hamartomatous polyps, increased melanin deposits, small bowel polyposis, and a family history of the syndrome.
3. Manifestations of the disease may first been countered by the dental professional during routine examination by the presence of melanotic pigmented spots in the oral cavity.

EPIDEMIOLOGY

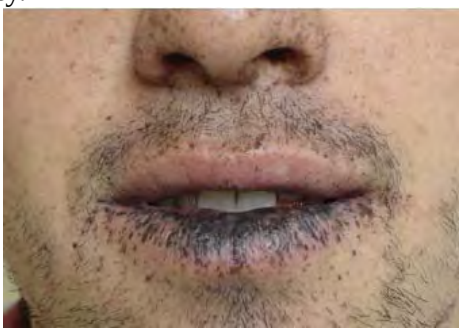
1. Jan Peutz + Harold Joseph Jeghers => Peutz Jeghers syndrome
2. PJS is, after juvenile polyposis, the most common hamartomatous syndrome among hereditary gastrointestinal polyposis syndromes.
3. Its prevalence is ap proximately1 in 200,000, with no evident gender or racial predilection.

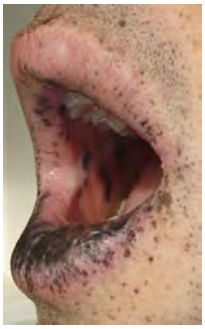
ETIOLOGY

1. Inheritable variations in the serine/threonine kinase gene (STK11/LKB1) are responsible for the pathogenesis of most cases of PJS. (tumor suppressor gene)
2. The germline mutation is identified in 30% to 80% of PJS patients.
3. The type of mutation and where it occurs within the gene sequence correlates with onset of PJS symptoms and its association with malignancies.

CLINICAL PRESENTATION

1. Hamartomatous polyps in the small intestine.(jejunum)
   A. usually appear during the first decade of life, and become symptomatic between the ages of 10 and 30 years.
   B. Size -> intussusceptions and bowel obstruction, severe abdominal pain, and gastrointestinal and rectal bleeding with anemia.
2. Mucocutaneous hyperpigmentation
   A. more than 90% of the patients with PJS.
   B. appears at an early age and fading of pigmented spots tends to occur during puberty.





3. differential diagnosis

polyposis	A. Familial juvenile polyposis

	B. Hereditary mixed polyposis syndrome C. Cowden's syndrome D. Cronkhite-Canada syndrome E. Familial adenomatous polyposis (Gardner's syndrome) F. Bannayan-Ruvalcaba-Riley syndrome G. Basal Cell Nevus syndrome (Gorlin syndrome)
multifocal pigmented lesions	A. Leopard syndrome B. Laugier-Hunziker syndrome C. Carney complex D. Cowden's syndrome

DIAGNOSIS

1. Three or more histologically confirmed benign hamartomatous polyps. (a connective tissue core infiltrated by smooth muscle, presence of goblet and paneth cells, and smooth muscle strands within the stroma, muscular core extends into the superficial epithelial layer forming a tree-like framework.(arborization))
2. Any number of hamartomatous polyps with a family history of PJS.
3. Mucocutaneous pigmentation with a family history of PJS.
4. Any number of hamartomatous polyps and mucocutaneous pigmentation.

MEDICAL MANAGEMENT

1. Surveillance and treatment of the hamartomatous polyps.
2. Conservative treatment, early screening and cancer detection in patients diagnosed with PJS.
3. MRI, testicular examinations, mammograms, Pap smears, and transvaginal ultrasound.
4. Chemoprevention of polyps with nonsteroidal anti-inflammatory drugs(NSAIDs) has been advocated for patients with familial adenomatous polyposis(FAP), but has not been recommended for other polyposis syndromes.
5. NSAIDs (celecoxib, sulindac): regression of existing adenomatous polyps, suppression of new polyps, reducing the incidence of colorectal cancer in FAP patients.
[celecoxib-- COX-2 inhibitor: ulceration and perforation of the gastric mucosa, increased risk of cardiovascular side effects.]
6. No standard treatment for mucocutaneous pigmentation.
   => Q-switched ruby laser (QSRL)

PROGNOSIS

1. Intussusception involving the small intestine is the most frequent complication of PJS.
   => Repeated surgical removal→ short bowel syndrome (SBS)
2. Intussusception of large hamartomatous polyps => acute upper gastrointestinal bleeding and chronic fecal blood loss => Iron-deficiency anemia => supplemental iron
3. Individuals affected with PJS are at increased risk for a wide variety of cancers. (gastrointestinal tract, lung, prostate, breast, and female reproductive organs) => mutated STK11/LKB1 gene
4. PJS is related to the increased risk of malignancy development.

ORAL MANIFESTATIONS

1. Extra- and intraoral pigmentation; on the vermilion border of the lips, labial mucosa, palate, and tongue; from 1 to 5mm in size.
2. Pigmentation may be noted at birth but usually develops early in childhood.
3. Oral lesions may be present before the onset of gastrointestinal disease.

4. Severe malabsorption => atrophic glossitis => taste sensation and food intake

DENTAL MANAGEMENT

1. Preoperative considerations
   a. Thorough review of medical history
   b. If known history of PJS
   c. Thorough extraoral and intraoral examination
   d. Evaluate hemoglobin, hematocrit, platelet count, and electrolytes.
2. Postoperative considerations: postoperative bleeding, Periodic evaluation, proper oral hygiene and balanced diet.
3. Iron-deficiency anemia and SBS => avoid long dental appointments and multiple procedures
4. Hemoglobin and hematocrit levels reach below 10g/dL and 30% => increased risk for intraoperative bleeding and decreased wound healing
5. When hemoglobin levels are between 6.0 and 8.0g/dL => transfusion (erythropoietin and iron)

6. PJS patients with SBS could have moderate to severe micronutrient deficiency. (essential fatty acids=> wound healing and platelet function)

CONCLUSION

Oral health care providers may play a significant role in detection and surveillance of PJS.

題號	題目
1	有關Peutz Jeghers syndrome，下列敘述何者錯誤? (A) 為一隱性體染色體疾病 (B) 有hamartomatous polyps (C) 病患較容易得到cancer (D) 有mucocutaneous hyperpigmentation
答案(A)	出處：Oral & Maxillofacial Pathology p.653
題號	題目
2	下列哪一種病患不會有pigmentation? (A) Addison's disease (B) Cushing's syndrome (C) Neurofibromatosis (D) Peutz Jeghers syndrome
答案(B)	出處：Oral & Maxillofacial Pathology