

A 25-hour-old full-term boy in the nursery is vomiting. His mother had attempted to breastfeed but stopped when he developed "bright green" vomiting. The infant has not yet had a bowel movement. Maternal prenatal screening was positive for a copy of the CFTR Δ F508 mutation. His paternal uncle had cystic fibrosis and died of respiratory failure. The boy's temperature is 36.7 C (98 F), blood pressure is 80/50 mm Hg, pulse is 154/min, and respirations are 46/min. Examination shows a crying infant with a markedly distended abdomen. Rectal examination shows no stool in the rectal vault. Feeds are held and a nasogastric tube is placed for decompression. X-rays of the abdomen show markedly dilated loops of bowel with no rectal air and no free air. Which of the following is the most appropriate next step in management of this patient?

- ☐ A. Anorectal manometry
- ☐ B. Computed tomography of the abdomen
- ☐ C. Contrast enema
- ☐ D. Rectal biopsy
- ☐ E. Serum lipase
- ☐ F. Surgery
- ☐ G. Sweat chloride testing

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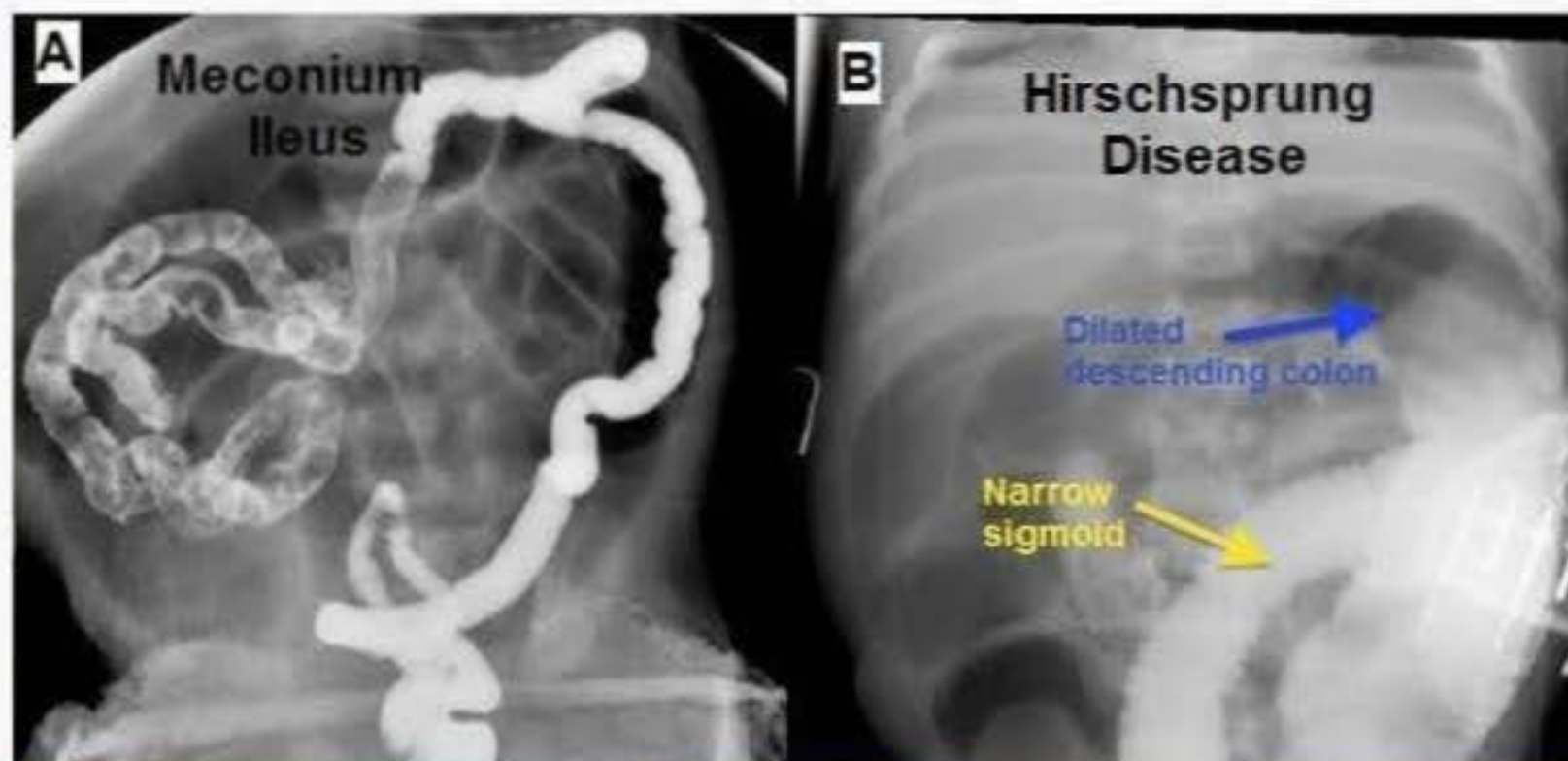
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- ☐ A. Anorectal manometry [4%]
- ☐ B. Computed tomography of the abdomen [4%]
- ☒ C. Contrast enema [41%]
- ☐ D. Rectal biopsy [6%]
- ☐ E. Serum lipase [1%]
- ☐ F. Surgery [19%]
- ☐ G. Sweat chloride testing [25%]

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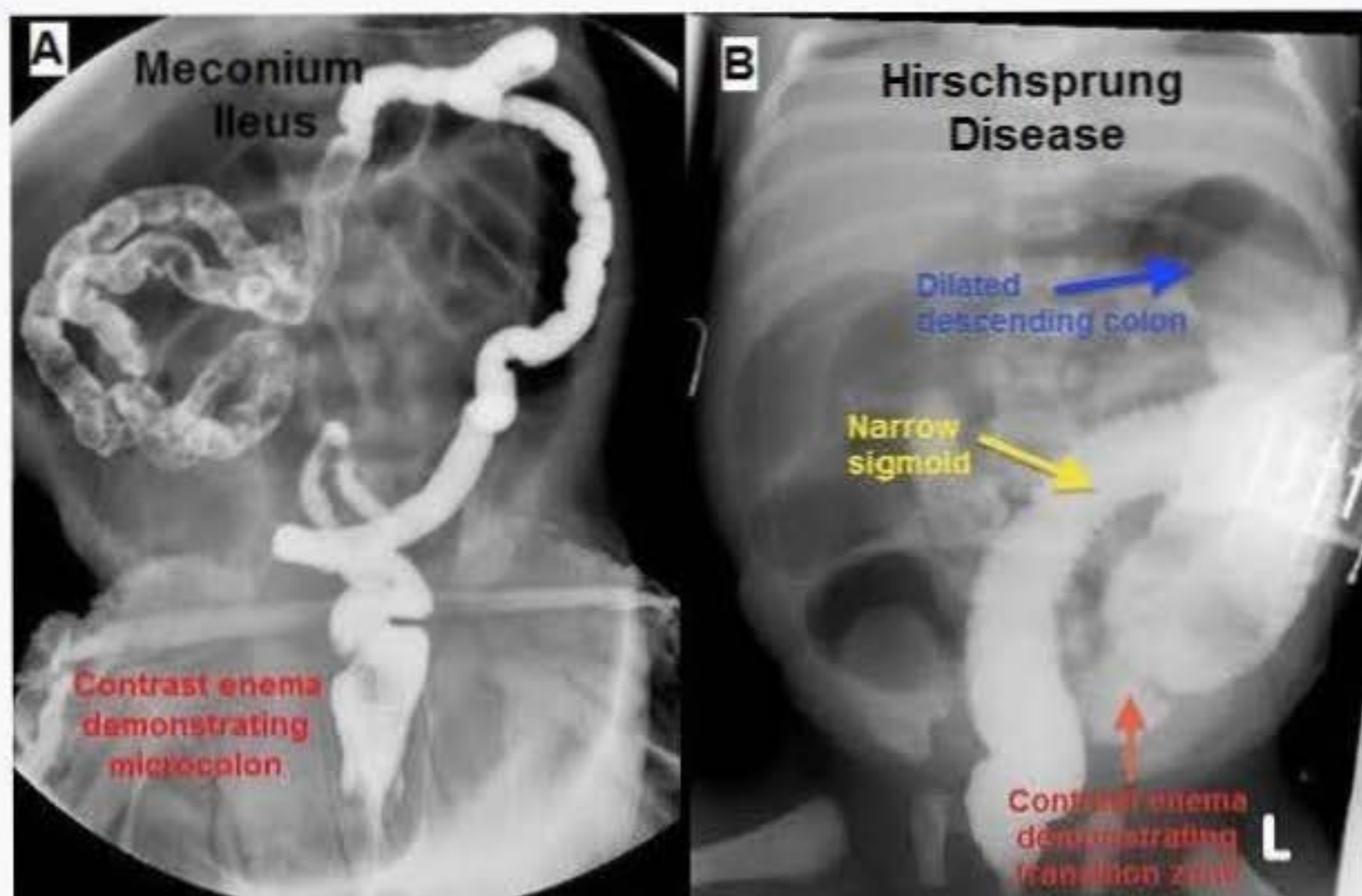
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Explanation:

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Bilious emesis in the neonate is an ominous sign of intestinal obstruction and requires **immediate workup**. Imaging should be performed in stable patients to delineate the level of obstruction and identify complications such as perforation. This patient is stable based on age-appropriate vital signs and lack of hematemesis. Because nearly all full-term infants pass meconium within 24-48 hours of life, distal intestinal obstruction should be suspected. Abdominal x-ray is always the first step as it can identify pneumoperitoneum from perforated bowel that would require emergency surgery (**Choice F**). After pneumoperitoneum has been excluded, water-soluble **contrast enema** is the best next step in management.

The appearance of a **microcolon** (image A above) on contrast enema should raise concern for **meconium ileus**. The underused, contracted colon is a result of **viscous meconium** accumulation and obstruction in the **terminal ileum**. Administration of hyperosmolar enema (eg, Gastrografin) can potentially break up the inspissated meconium and dissolve the obstruction. Surgery is required if therapeutic enema is unsuccessful.

Meconium ileus is virtually pathognomonic for **cystic fibrosis (CF)**. CF is the most common autosomal recessive disease in white populations, and up to 20% of newborns with CF present with meconium ileus. **ΔF508** is the predominant mutation responsible for

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Meconium ileus is virtually pathognomonic for **cystic fibrosis** (CF). CF is the most common autosomal recessive disease in white populations, and up to 20% of newborns with CF present with meconium ileus. **ΔF508** is the predominant mutation responsible for defective chloride transport and tenacious secretions in multiple organs.

(Choices A and D) If contrast enema demonstrates a transition zone between a narrow rectosigmoid and a dilated megacolon (image B above), workup for congenital aganglionic megacolon (Hirschsprung disease) should be pursued. Rectal biopsy should be performed; the absence of ganglion cells would confirm the diagnosis. Anorectal manometry may be necessary if results of rectal biopsy are equivocal, but neither of these tests is warranted for this infant.

(Choice B) Computed tomography of the abdomen should not be the initial diagnostic test for obstruction in neonates due to the significant amount of radiation exposure compared to contrast studies.

(Choice E) Pancreatitis is extremely uncommon in neonates. Although pancreatitis occurs in 10% of patients with CF, it typically occurs during late adolescence or early adulthood. Serum lipase would be low yield, and the focus should be on diagnosing gastrointestinal obstruction.

(Choice G) All patients with meconium ileus require sweat chloride testing to confirm diagnosis of CF. However, the priority for this patient is to identify and treat the acute obstruction.

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Educational objective:

Neonatal bilious emesis signifies bowel obstruction and requires immediate x-ray to evaluate the need for emergency surgery or further diagnostic studies. Contrast studies should be performed in stable patients to determine the level of obstruction.

Media Exhibit

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Media Exhibit

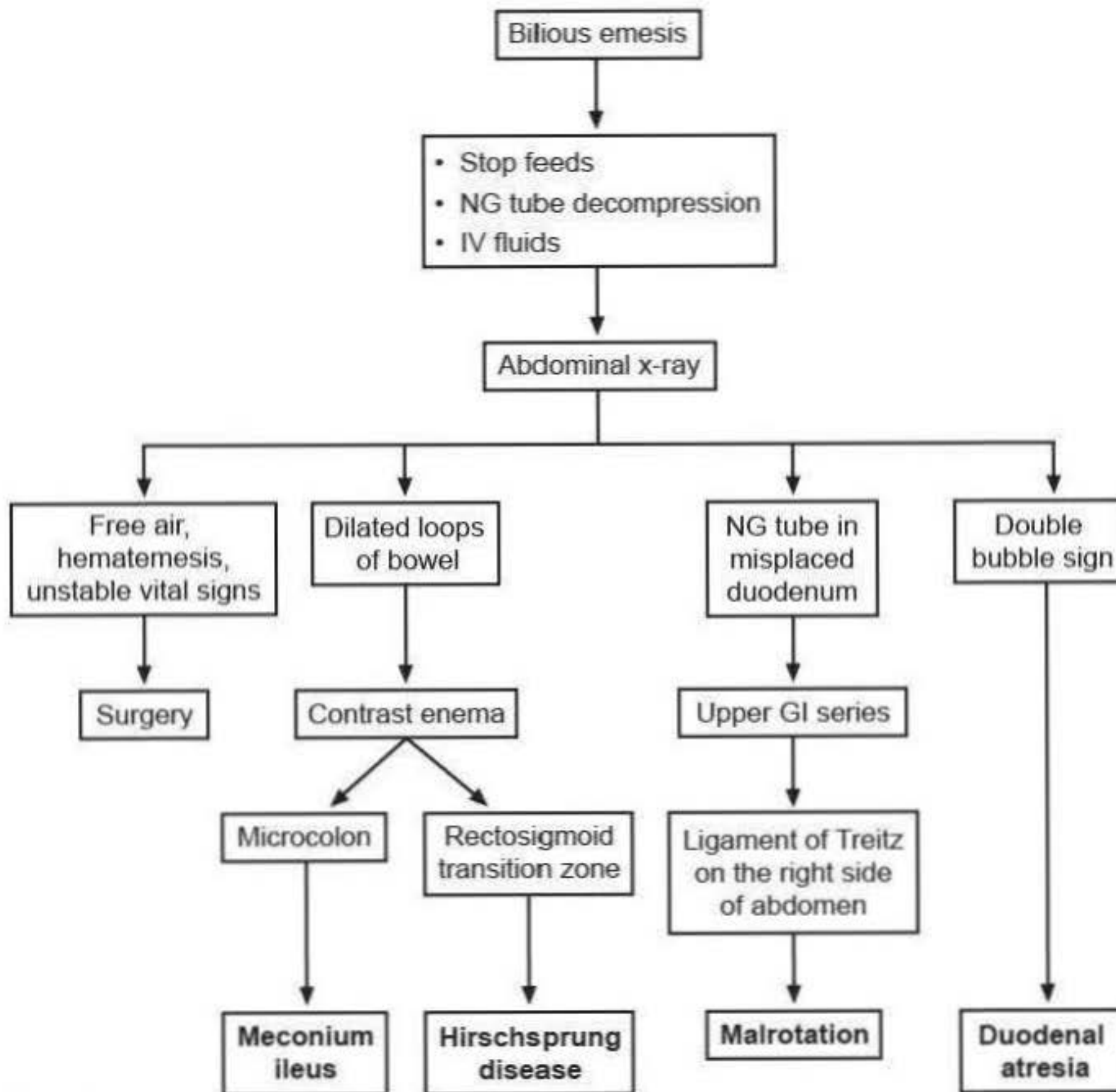
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Media Exhibit

on of neonatal bilious emesis

Evaluation of bilious emesis in the neonate



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