STÉNOSE DU PYLORE

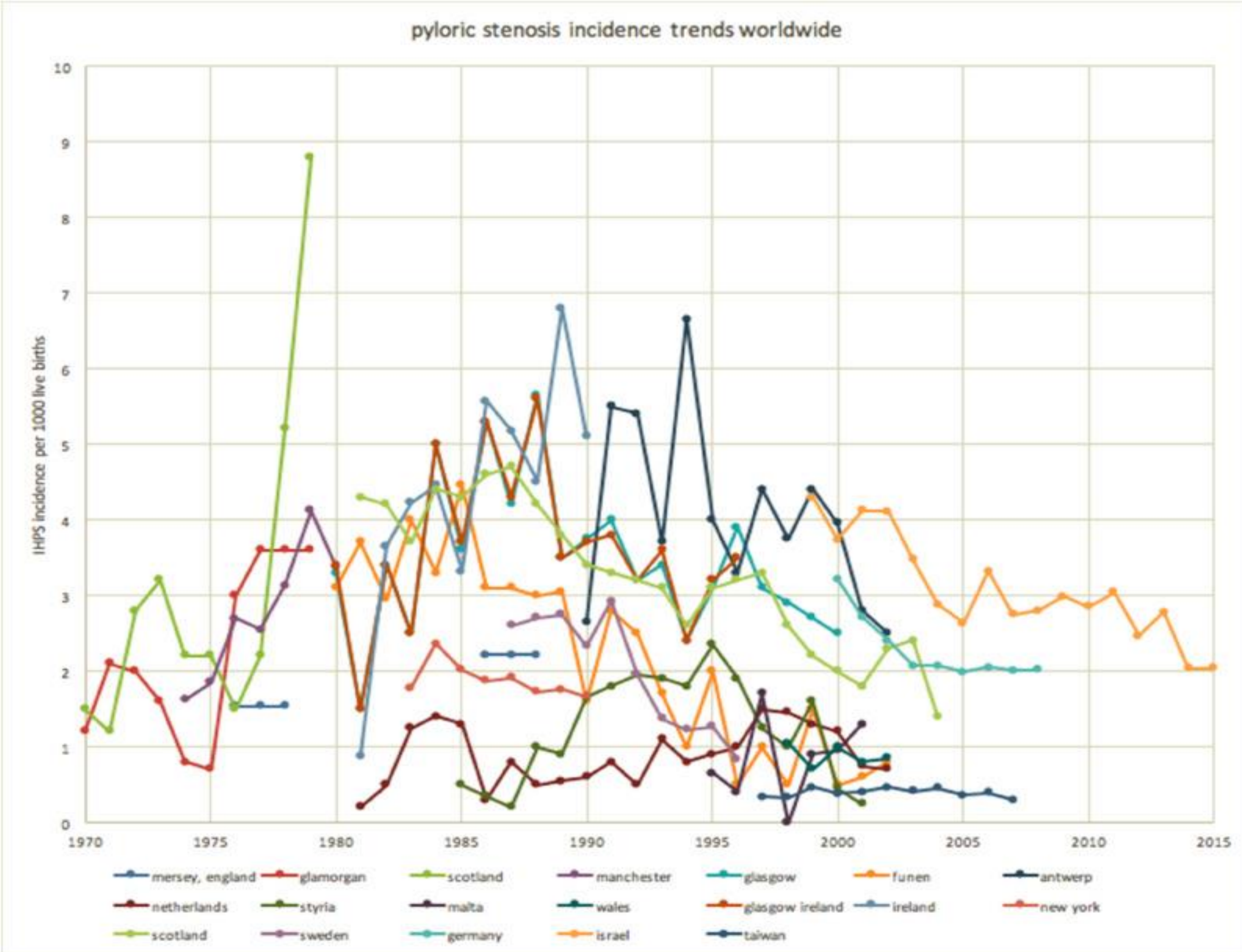
Guillaume LOPIN
Urgences Pédiatriques
CHU de Toulouse
JUP, le 4 octobre 2018

- Je n’ai pas de conflit d’intérêt

« Epaississement des couches musculaires pyloriques par hypertrophie et hyperplasie des fibres musculaires entrainant un obstacle à la sortie de l’estomac »

Epidémiologie

- **1 à 4 pour 1000** naissances vivantes
- Grandes **variations** régionales
- Blancs > Noirs > Asiatiques



pyloric stenosis incidence trends worldwide
IHPS incidence per 1000 live births
0
1
2
3
4
5
6
7
8
9
10
1970
1975
1980
1985
1990
1995
2000
2005
2010
2015
mersey, england
glamorgan
scotland
manchester
glasgow
funen
antwerp
netherlands
styria
malta
wales
glasgow ireland
ireland
new york
scotland
sweden
germany
israel
taiwan

Epidémiologie

- Sex ratio de **4:1 à 6:1**
- Variation saisonnière : automne et printemps
- **Survie 100%**

Epidémiologie

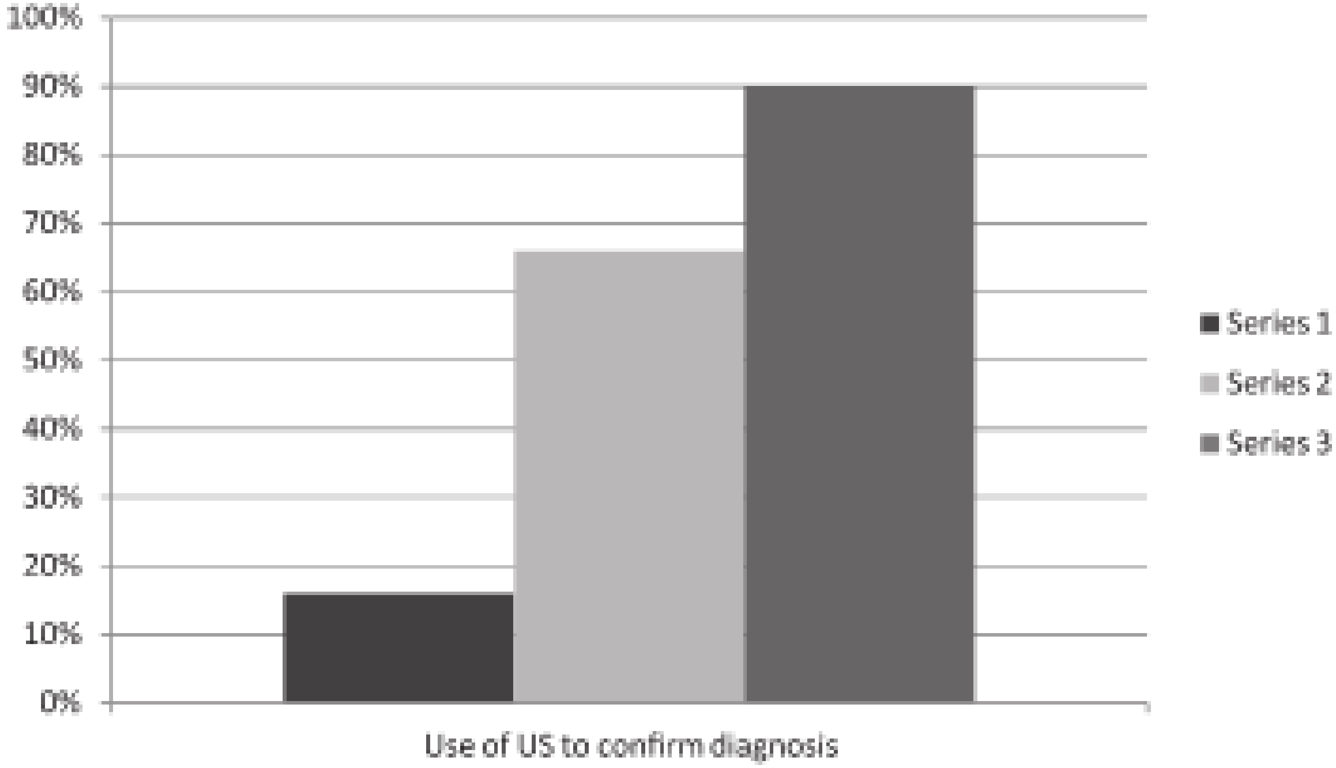
- Début des symptômes **entre 3 et 6 semaines**
- 95% des cas entre 3 semaines et 3 mois
- Exceptionnel après 12 semaines

Epidémiologie

- Age au moment du diagnostic en baisse :
  - âge moyen 5,4 semaines en 1975 vs 3,4 semaines en 1995
  - Les mé
  - Les pa
  - Accès

	1973–1988 N = 1890 No. (%)	1989–1997 N = 1248 No. (%)	1998–2008 N = 658 No. (%)	Total N = 3796 No. (%)
Gender				
Boy	1576 (83.4)	1038 (83.2)	565 (85.9)	3179 (83.7)
Girl	314 (16.6)	210 (16.8)	93 (14.1)	617 (16.3)
Twin				
Born as twin	80 (4.2)	62 (5.0)	46 (7.0)	188 (5.0)
Not twin	1810 (95.8)	1186 (95.0)	612 (93.0)	3608 (95.0)
Age at diagnosis (weeks)				
1–2	39 (2.1)	26 (2.1)	21 (3.2)	86 (2.3)
3–4	374 (19.8)	303 (24.3)	198 (30.1)	875 (23.1)
5–6	697 (36.9)	468 (37.5)	229 (34.8)	1394 (36.7)
7–8	378 (20.0)	228 (18.3)	118 (17.9)	724 (19.1)
9–10	211 (11.2)	112 (9.0)	52 (7.9)	375 (9.9)
11–12	81 (4.3)	55 (4.4)	28 (4.3)	164 (4.3)
13–14	40 (2.1)	26 (2.1)	5 (0.8)	71 (1.2)
≥15	70 (3.7)	30 (2.6)	7 (1.1)	107 (2.8)

Epidémiologie



1984-1989

1990-1995

2004-2010

Fig. 4 Frequency of use of ultrasound (US) to confirm diagnosis.

Nicole D Taylor, Daniel T Cass, Andrew JA Holland, Infantile hypertrophic pyloric stenosis : Has anything changed?, Journal of paediatrics and child health, 49 (2013) 33-37

Etiologie

Théorie Neurogène

Immaturité innervation gastrique

↓

Dyssinergie antro-pylorique

↓

Contractions désordonnées butant sur spasme pylorique

↓

Hypertrophie musculaire pylorique

Théorie Hormonale

Stress à la naissance

↓

Sécrétion de gastrine

↓

Contraction et spasme du pylore

↓

Hypertrophie musculaire

↓

Distension gastrique

Théorie Génétique

Familial Aggregation and Heritability of Pyloric Stenosis

Camilla Krogh, MD
Thea K. Fischer, DMSc
Line Skotte, MSc
Robert J. Biggar, MD
Nina Øyen, DMSc
Axel Skytthe, PhD
Sanne Goertz, MSc
Kaare Christensen, DMSc
Jan Wohlfahrt, DMSc
Mads Melbye, DMSc

Context Pyloric stenosis is the most common condition requiring surgery in the first months of life. Case reports have suggested familial aggregation, but to what extent this is caused by common environment or inheritance is unknown.

Objectives To investigate familial aggregation of pyloric stenosis from monozygotic twins to fourth-generation relatives according to sex and maternal and paternal contributions and to estimate disease heritability.

Design, Setting, and Patients Population-based cohort study of 1 999 738 children born in Denmark between 1977 and 2008 and followed up for the first year of life, during which 3362 children had surgery for pyloric stenosis.

Main Outcome Measure Familial aggregation of pyloric stenosis, evaluated by rate ratios.

Results The incidence rate (per 1000 person-years) of pyloric stenosis in the first year of life was 1.8 for singletons and 3.1 for twins. The rate ratios of pyloric stenosis were 182 (95% confidence interval [CI], 70.7-467) for monozygotic twins, 29.4 (95% CI, 9.45-91.5) for dizygotic twins, 18.5 (95% CI, 13.7-25.1) for siblings, 4.99 (95% CI, 2.59-9.65) for half-siblings, 3.06 (95% CI, 2.10-4.44) for cousins, and 1.60 (95% CI, 0.51-4.99) for half-cousins. We found no difference in rate ratios for maternal and paternal relatives of children with pyloric stenosis and no difference according to sex of cohort member or sex of relative. The heritability of pyloric stenosis was 87%.

Conclusion Pyloric stenosis in Danish children shows strong familial aggregation and heritability.

JAMA. 2010;303(23):2393-2399 www.jama.com

PYLORIC STENOSIS IS A SEVERE AND potentially fatal condition in which apparently healthy infants, typically 2 to 8 weeks old,[1-5] develop an inability to pass food from the stomach into the duodenum. The condition is characterized by hypertrophy of the pylorus smooth circular muscle layer. Mortality was high

Théorie Génétique

- **Agrégation familiale** significative
  - Jumeaux monozigotes : risque x200
  - Jumeaux dizigotes : risque x20
  - Contribution Mère = Père (environnement utérin non impliqué)

- **Héritabilité 87%**

- **Plusieurs loci** identifiés
  - MBNL1
  - NKX2-5
  - Groupe de gènes de l'APOA1 : associations entre faible taux cholestérol plasmatique / risque sténose du pylore

Krogh C, Fischer TK, Skotte L, et al. Familial aggregation and heritability of pyloric stenosis. JAMA 2010; 303:2393

Facteurs de risque

- Garçon
  - Environ 80%
  - Sex ratio 4:1
- Premier né
  - 30 à 40% des cas
  - Risque x1,5

Facteurs de risque

- Antécédents familiaux
- Prématurité
- Mère jeune
- Césarienne
- Niveau socio-économique bas

	Cases (n = 3608)[a] No. (%)	Controls (n = 17588)[a] No. (%)	Unadjusted OR (95% CI)	Adjusted OR (95% CI)
Maternal age (years)				
<20	157 (4.4)	539 (3.1)	1.53 (1.27–1.85)	1.42 (1.17–1.73)
20–24	967 (26.8)	3723 (21.2)	1.36 (1.23–1.49)	1.33 (1.21–1.47)
25–29	1228 (32)	6387 (36.3)	1	1
30–34	827 (32)	4711 (36.3)	0.91 (0.82–1.00)	0.94 (0.85–1.03)
≥35	429 (11.9)	2228 (12.7)	0.99 (0.87–1.11)	0.98 (0.86–1.12)

Svenningsson A, Svensson T, Akre O, Nordenskjöld A, Maternal and pregnancy characteritics and risk of infantile hypertrophic pyloric stenosis, Journal of Pediatric surgery, Aout 2014

Facteurs environnementaux

- **Tabagisme** maternel pendant la grossesse : risque x1,5 à 2

- A

	High incidence 1982–1988 Cases n = 1114[a] Controls n = 5447[a] OR (95% CI)	Decreasing incidence 1989–1997 Cases n = 1186[a] Controls n = 5769[a] OR (95% CI)	Low incidence 1998–2008 Cases n = 612[a] Controls n = 2973[a] OR (95% CI)	P-value[b]
Maternal smoking[c] (cigarettes daily)[d]				
None	1	1	1	
1–9	1.48 (1.20–1.82)	1.66 (1.30–2.12)	1.98 (1.43–2.74)	<0.001
≥10	1.56 (1.22–1.98)	2.15 (1.62–2.87)	2.05 (1.21–3.49)	<0.001

diatrics 2012; 130:e943

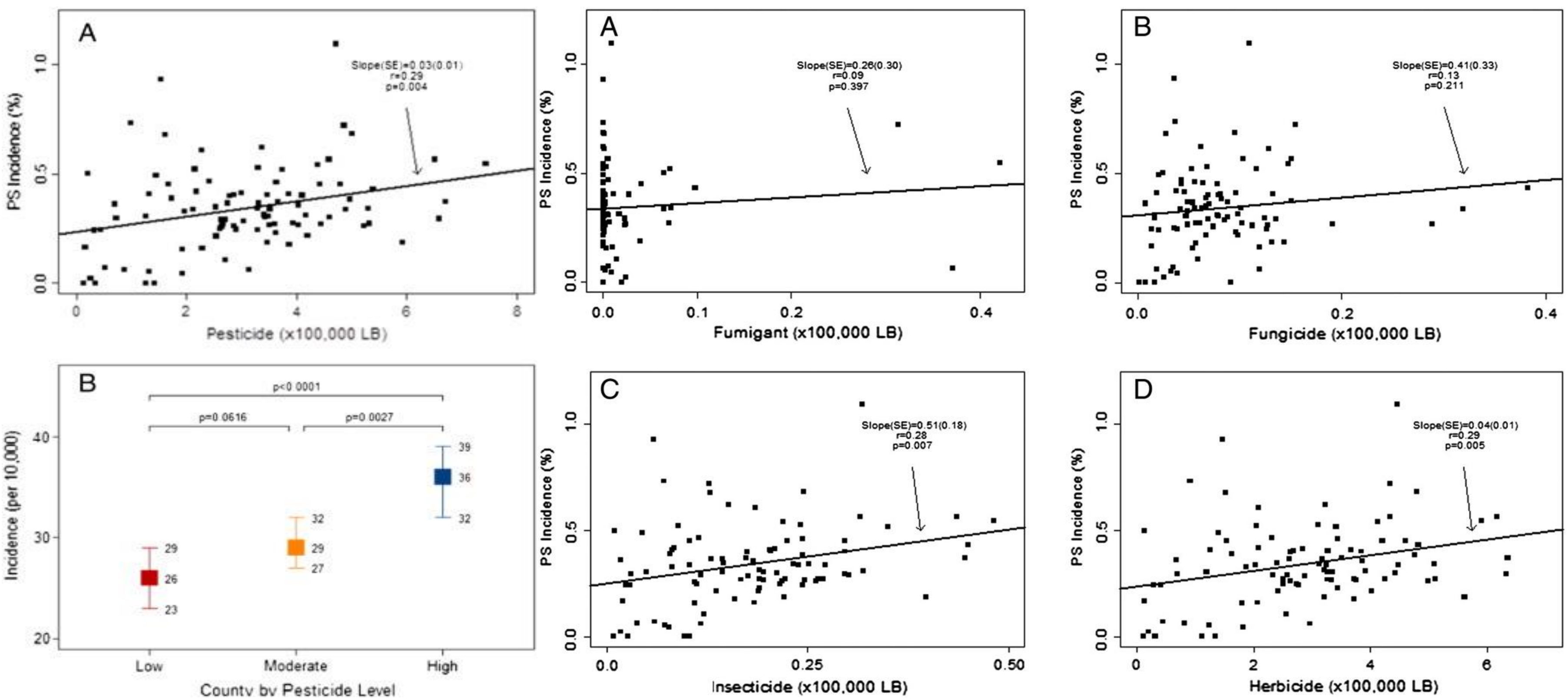
McAteer JP, Ledbetter DJ, Goldin AB. Role of bottle feeding in the etiology of hypertrophic pyloric stenosis. JAMA Pediatr 2013; 167:1143

Facteurs environnementaux

- **Antibiotiques**
  - Erythromycine et Azithromycine : risque augmenté
    - Nouveau-né de moins de 2 semaines
    - Macrolides chez mères en fin de grossesse ou pendant l'allaitement

Facteurs de risque environnementaux

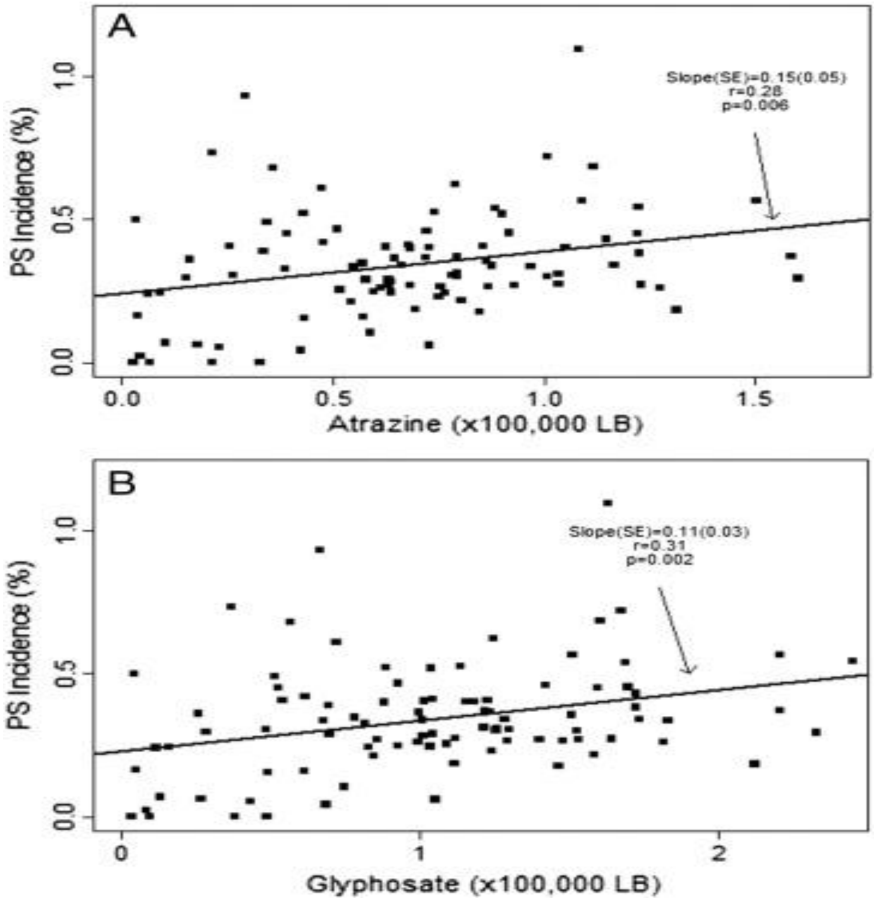
- Pesticides



Troy A. Markel, Cathy Proctor, Jun Ying, Paul D. Winchester, Environnemental pesticides increase the risk of developing hypertrophic stenosis, Journal of Pediatric Surgery, Vol 50, Issue 8, Aout 2015, p. 1283-1288

Facteurs de risque environnementaux

- Pesticides



Troy A. Markel, Cathy Proctor, Jun Ying, Paul D. Winchester, Environnemental pesticides increase the risk of developing hypertrophic stenosis, Journal of Pediatric Surgery, Vol 50, Issue 8, Aout 2015, p. 1283-1288

Facteurs environnementaux

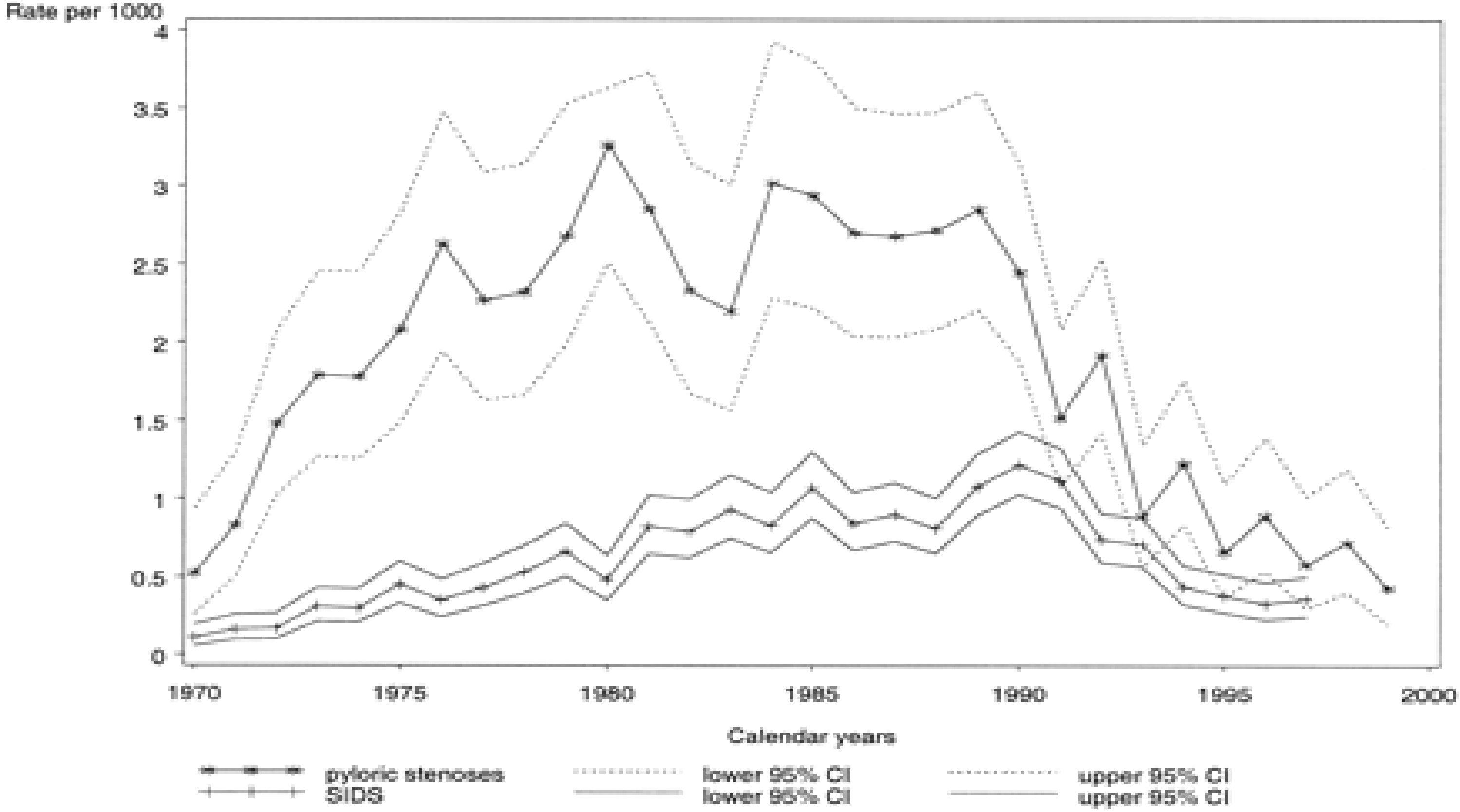
- Non prouvé :
  - Rôle des infections ?
  - Rôle d'hélicobacter pylori ?
  - Rôle du déficit en folate ?
  - Position de couchage ?

Facteurs environnementaux

- Position de couchage

Svenningsson A, Svensson T, Akre O, Nordenskjöld A, Maternal and pregnancy characteritics and risk of infantile hypertrophic pyloric stenosis, Journal of Pediatric surgery, Aout 2014

Facteurs environnementaux



De Laffolie J, Turial S, Heckmann M, Zimmer KP, Schier F, Decline in infantile pyloric stenosis in Germany in 2000-2008, Pediatrics 2012
Lisonkova S, Joseph KS, Similarities and differences in the epidemiology of pyloric stenosis and SIDS, Matern Child Health, Sept 2014
Persson S, EkbomA, Granath F, NordenskjoldA. Parallel incidences of sudden infant death syndrome and infantile hypertrophic pyloric stenosis : a common cause? Pediatrics, 2001;108:E70

Manifestations cliniques

- Vomissements
  - Post-prandiaux systématiques
  - D'aggravation progressive
  - Non inquiétant en volume et
    en fréquence au début
  - Puis explosifs, en jet et volumineux
  - Parfois tardifs
  - De lait caillé

  - Non bilieux
    - 1,4% de vomissements bilieux sur une série de 354 sténoses du pylore

Piroutek MJ, Brown L, Throp AW, Bilious vomiting does not rule out infantile hypertrophic pyloric stenosis, Clin Pediatr (Phila), Mars 2012)

  - Rarement hématémèse (sur oesophagite)

La répétition de vomissements non bilieux,

en particulier si consultations multiples,

chez un NRS de 3 à 6 semaines doit conduire à

demander une échographie du pylore

Manifestations cliniques

- Raréfaction des selles
- Appétit conservé voire nourrisson affamé

Manifestations cliniques

- Stagnation voire perte de poids

- Déshydratation extra-cellulaire
  - Cernes
  - Pli cutané
  - Oligurie
  - Dépression FA

Dénutrition et déshydratation de moins en moins fréquents (diagnostic de + en + précoce)

Manifestations cliniques

Conséquences de l'alcalose métabolique

Non spécifiques et tardives++

- **Dépression ventilatoire**

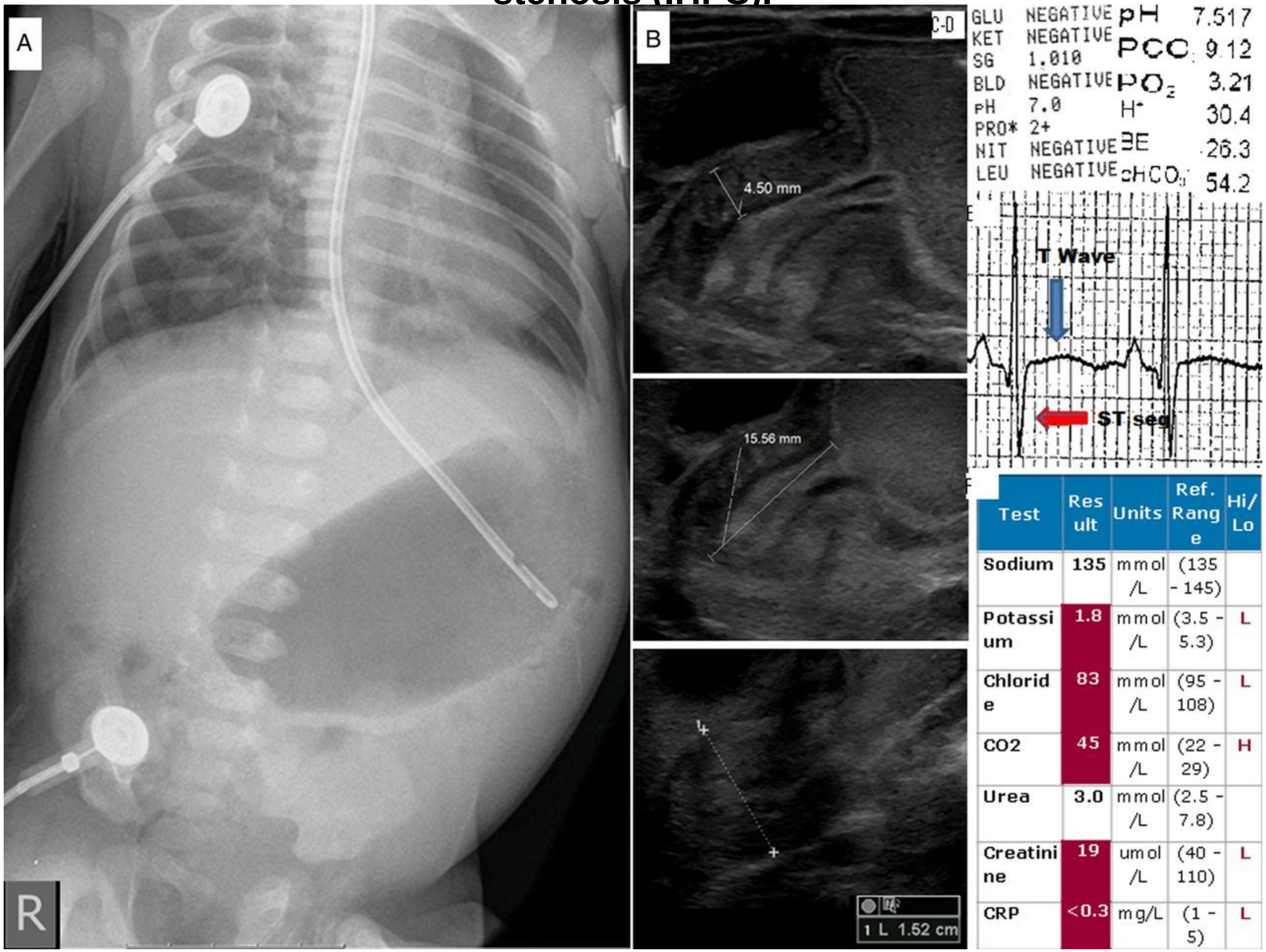
→ hypoxie, atélectasies

- **Vasoconstriction** et diminution de la libération d'O2 par l'Hb

→ ischémie myocardique (tdr cardiaque)

→ et cérébrale (apathie, convulsions)

(A) Babygram, (B) abdominal ultrasound, (C) urinalysis, (D) capillary blood gas, (E) ECG and (F) serum electrolytes showing typical changes of advanced infantile hypertrophic pyloric stenosis (IHPS).



Test	Result	Units	Ref. Range	Hi/Lo
Sodium	135	mmol/L	(135 - 145)	
Potassium	1.8	mmol/L	(3.5 - 5.3)	L
Chloride	83	mmol/L	(95 - 108)	L
CO2	45	mmol/L	(22 - 29)	H
Urea	3.0	mmol/L	(2.5 - 7.8)	
Creatinine	19	umol/L	(40 - 110)	L
CRP	<0.3	mg/L	(1 - 5)	L

=68mmHg
=24mmHg

Ramnik V Patel et al. BMJ Case Reports 2013;2013:bcr-2013-201435

Manifestations cliniques

- Prématuré
  - Age chronologique plus avancé
  - Vomissements parfois non en jet
  - Pleurs de faim parfois absents

Clinical diagnosis of pyloric stenosis: a declining art

Joseph Macdessi, R Kim Oates

Abstract

***Objective*—To assess whether diagnostic imaging of pyloric stenosis has made a difference in rapidity of diagnosis, duration of pre-operative hydration, and length of stay in hospital.**

***Design*—Chart review of infants with confirmed**

techniques between 1980 and 1984, largely attributable to the use of ultrasound.[5] In both studies the authors emphasised that most of the pyloric stenoses could have been diagnosed clinically and that many of the imaging studies performed were not only unnecessary but also not cost effective. It has been recom-

Sydney University, Department of Paediatrics and Child Health, Children's Hospital, Camperdown NSW 2050, Australia
Joseph Macdessi, *resident medical officer*
R Kim Oates, *professor of paediatrics and child health*

Correspondence to: Professor Oates.

BMJ 1993;306:553-5

1967 found that a palpable pyloric mass was detected in 80% of cases.[6] From a study of 200 infants Stunden *et al* suggested that diagnosis by means of clinical methods alone is possible in 84% of cases.[7] Such patients should be operated on without undergoing imaging studies.[2 3 4]

In 1934 it was shown that pyloric stenosis could be confirmed by barium meal examination,[9] and in 1977 Teele and Smith first described the use of ultrasound in the diagnosis.[10] These techniques have been relied on increasingly to evaluate vomiting infants. Scharli *et al* noted a 23% rise in the use of barium meal examinations over the 55 years of their study,[4] while Breaux *et al* reported a 34% increase in the use of imaging

the records of 402 were available for study: 215 during 1974-7 (group A) and 187 during 1988-91 (group B).

Age, sex, and clinical presentation

The ratio of male to female infants in group A (3·8:1) was similar to that in group B (4·8:1). The average age of infants at presentation was 40 days in both groups, and 188 (87%) of those in group A and 168 (90%) of those in group B were aged between 2 weeks and 2 months on admission. The median duration of vomiting before admission was also similar in both groups, being seven days in group A and eight days in group B ($W=39799\cdot5$, $p=0\cdot34$ (95% confidence interval $-2\cdot0$ to $0\cdot0$)). Thirty nine infants (18%) in

Examen clinique

- Abdomen plat
- Indolore
- Diminution des BHA
- Olive pylorique
- Ondulations péristaltiques

L’olive pylorique

- Voussure en hypochondre D
- Sur le bord latéral du muscle droit
- Masse ferme qui roule sous les doigts
- A rechercher chez un enfant calme
- Immédiatement après administration G30%
- Plus facile après sondage gastrique

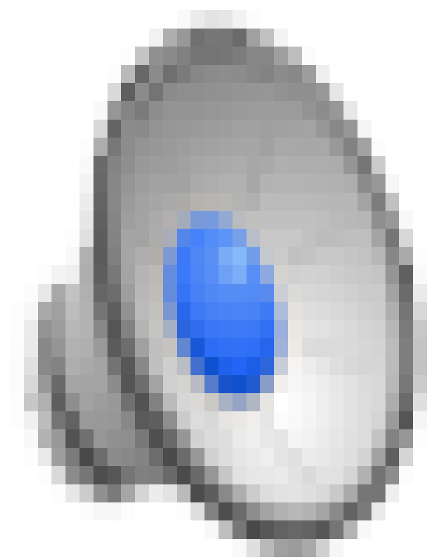
L'olive pylorique

- **Pathognomonique**
- **En pratique rarement palpé** (sauf sous AG)
- 87% (années 80) ➔ 48%
  - Echographie immédiate devant des symptômes évocateurs : perte habitude
  - NRS moins dénutris => Palpation moins facile

Bakal U, Sarac M, Aydin M, et al. Recent changes in the features of hypertrophic pyloric stenosis. Pediatr Int 2016; 58:369.
Macdessi J, Oates RK. Clinical diagnosis of pyloric stenosis: a declining art. BMJ 1993; 306:553.
Nicole D Taylor, Daniel T Cass, Andrew JA Holland, Infantile hypertrophic pyloric stenosis : Has anything changed?, Journal of paediatrics and child health, 49 (2013) 33-37

Ondulations péristaltiques

- Rarement visible (tardifs++, chez un NRS dénutri)
- Juste après un vomissement
- De gauche à droite



Biologie

- Alcalose hypochlorémique
  - pH>7,45
  - BE>+3
  - Cl-<98mmol/L
  - Chez un NRS vomisseur

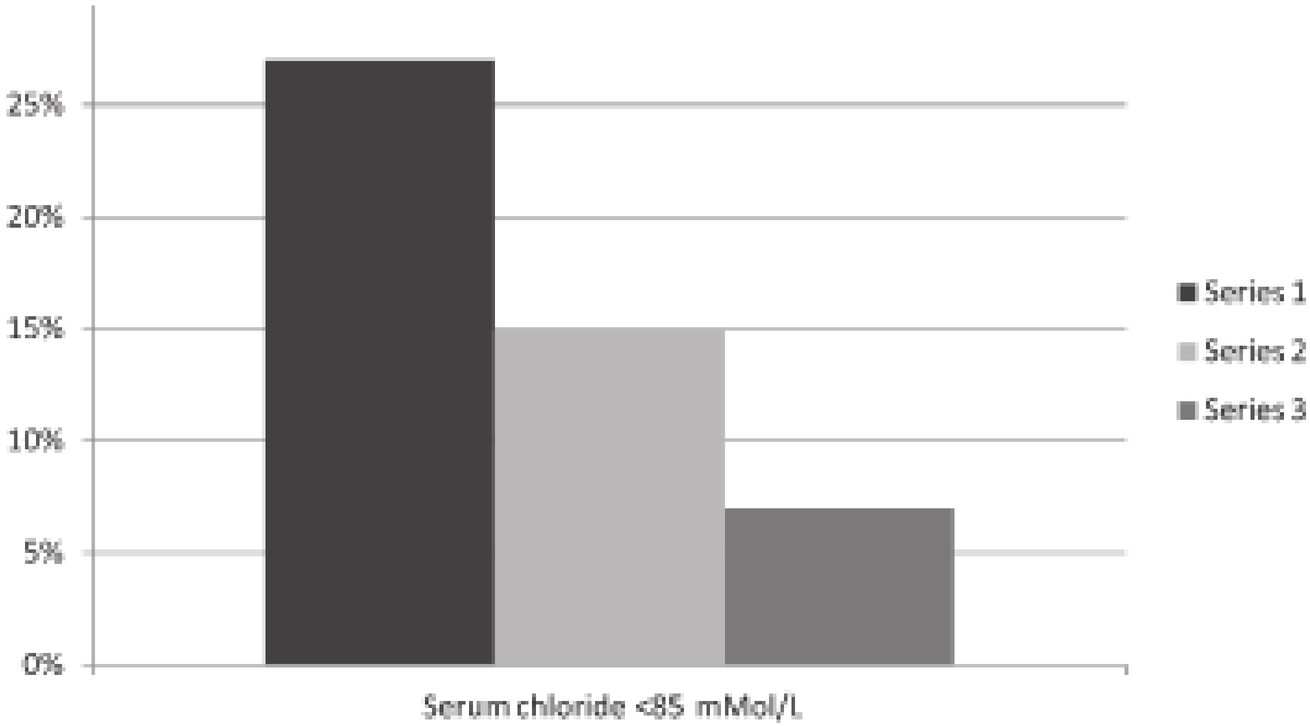
VPP=88%

Oakley EA, Barnett PL. Is acid base determination an accurate predictor of pyloric stenosis? J Paediatr Child Health 2000; 36:587

- Tardifs :
  - Acidurie paradoxale
  - Hypokaliémie
  - Dysnatrémie

- **BES et GDS le plus souvent normaux** : diagnostic précoce

Biologie



1984-1989

1990-1995

2004-2010

Fig. 3 Frequency of hypochloraemia.

Infantile hypertrophic pyloric stenosis : Has anything changed?
Nicole D Taylor, Daniel T Cass
Jounral of paediatrics and child Health, 2012

Biologie

- Hyperbilirubinémie : **sd ictéro-pylorique**
    - 14% des cas dans une série
    - Conjuguée
    - Résorption post-op
    - Manifestation précoce d'une maladie de Gilbert ?

Hua L, Shi D, Bishop PR, et al. The role of UGT1A1*28 mutation in jaundiced infants with hypertrophic pyloric stenosis. Pediatr Res 2005; 58:881

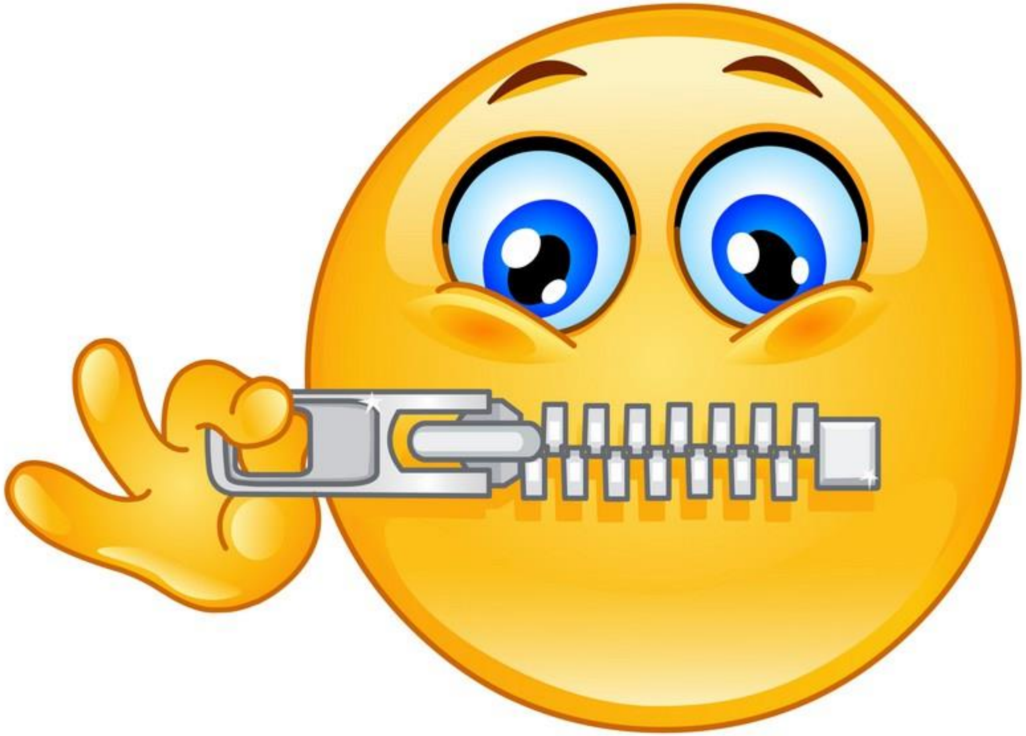
Diagnostics différentiels

- Occlusion intestinale : Mésentère commun, IIA, Hirschprung
- RGO
- GEA
- APLV
- Insuffisance surrénalienne
- Hépatopathies : Atrésie des VB, Maladies métaboliques

Imagerie :

Echographie++

Reste-t-il une place pour l’ASP et le TOGD?



PEC chirurgicale…

…**APRÈS CORRECTION DES TROUBLES HYDRO-ÉLECTROLYTIQUES**

Mise en condition

- A jeun
- Proclive
- SNG double courant type Salem (RP de contrôle)
- VVP
- NFS BES Urée Créatinine BH GDS TP TCA

Si signes de signes de gravité

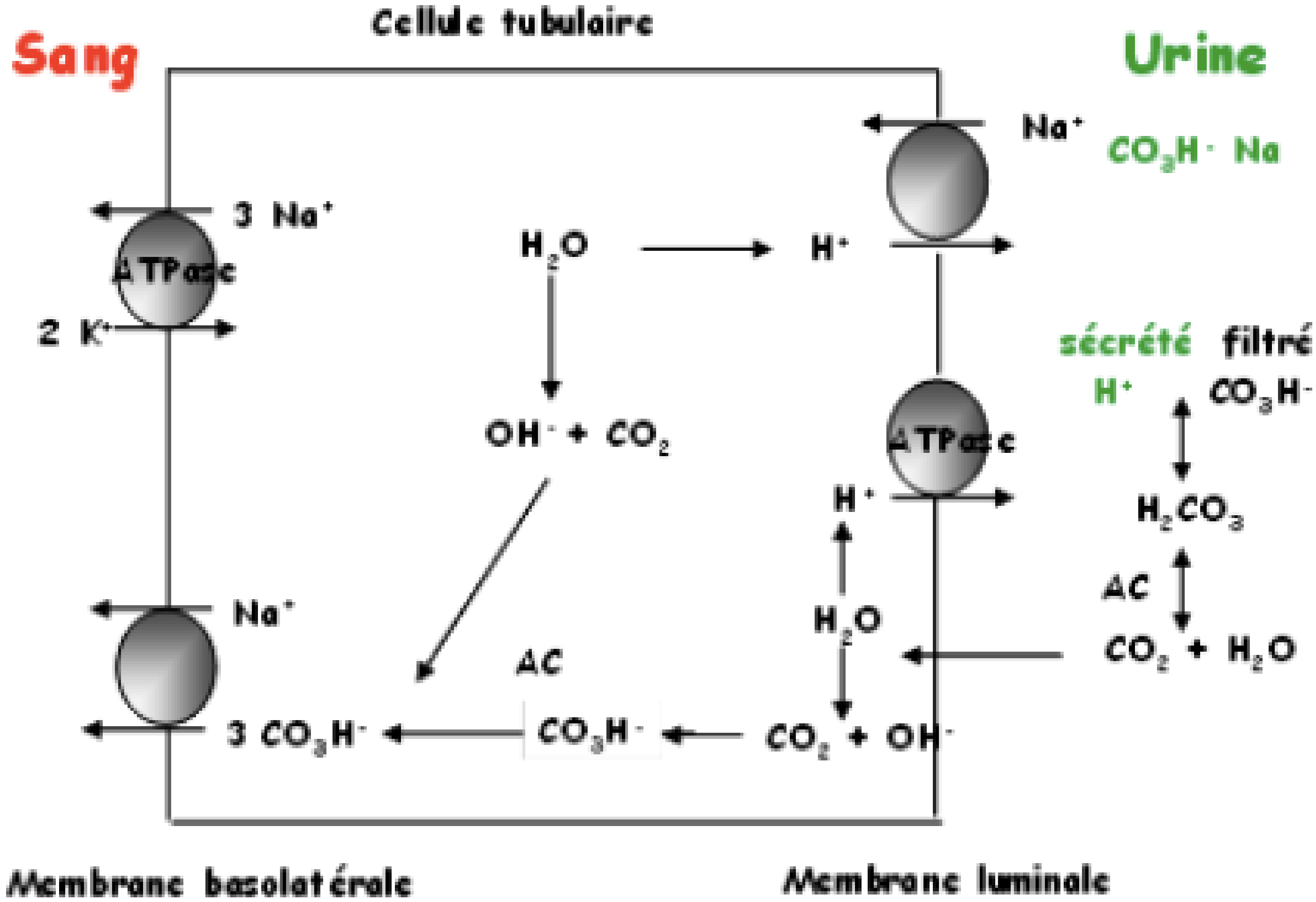
ALLO REA

- Choc hypovolémique
- RA>40
- Cl-<65 mmol/L

Si anomalies hydro-électrolytiques

- Déshydratation >10%
- AEG marquée
- Alcalose hypochlorémique
- Hypokaliémie

SEULS L'APPORT DE CHLORE ET LA CORRECTION DE L'HYPOVOLÉMIE PERMETTENT DE CORRIGER L'ALCALOSE, L'HYPOCL- ET L'HYPOK+


Cellule tubulaire
Sang
Urine
Na+
CO3H- Na
3 Na+
ATPase
2 K+
H2O
H+
sécrété
filtré
H+
CO3H-
OH- + CO2
ATPase
H+
H2CO3
Na+
H2O
AC
AC
CO2 + H2O
3 CO3H-
CO3H-
CO2 + OH-
Membrane basolatérale
Membrane luminale

Protocole CHU Toulouse

- A) **La 1ere**
  20 ml/kg de
- B) **Les 11 h**
  80 ml/kg av
  + 1 meq/kg d
  + 1 meq/kg d

zo

- C) **Si RA < :**
  Réhydratatio
  + 2 meq/kg N
  + 1 meq/Kg I
- **Sinon** repre

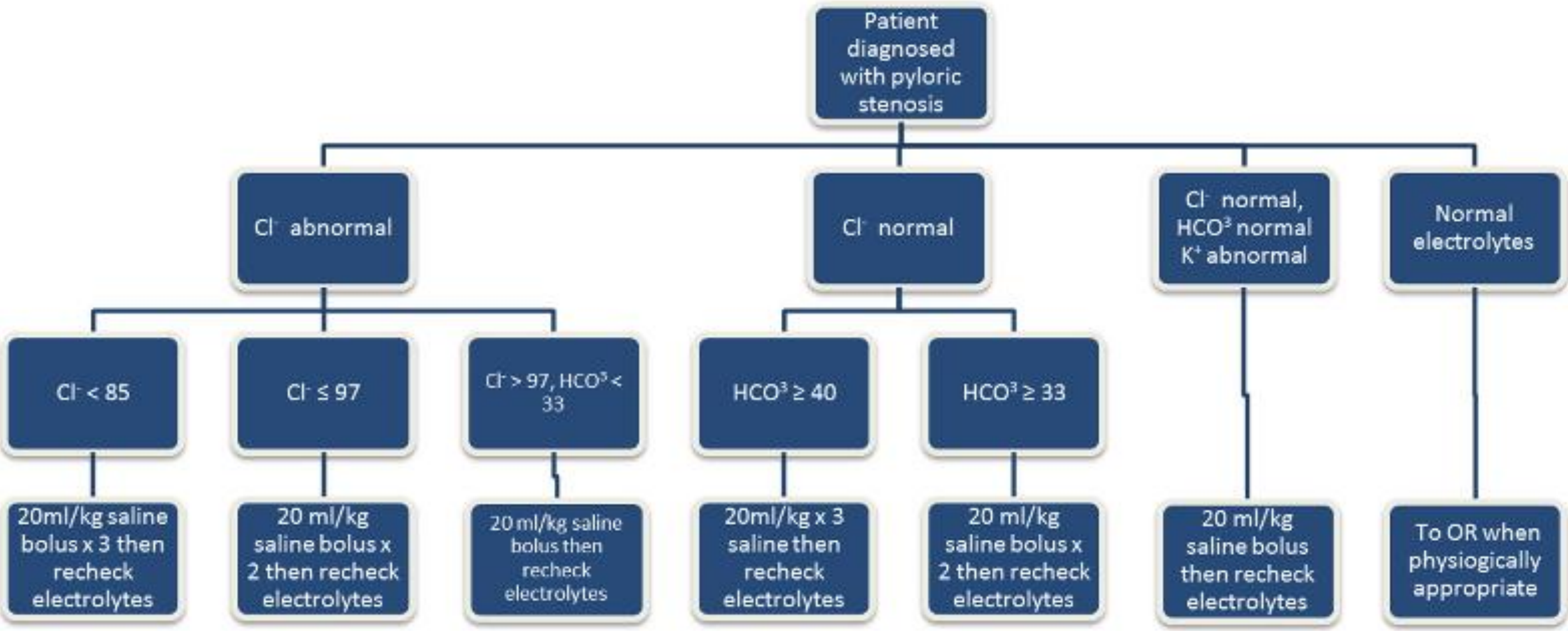


Fig. 4. Fluid resuscitation algorithm based on initial electrolyte values in hypertrophic pyloric stenosis.

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Optimizing fluid resuscitation in hypertrophic pyloric stenosis

Brian G.A. Dalton [a], Katherine W. Gonzalez [a], Sushanth R. Boda [a], Priscilla G. Thomas [a], Ashley K. Sherman [b], Shawn D. St. Peter [a,*]

[a] *Department of Pediatric Surgery, Children's Mercy Hospital, Kansas City, MO*
[b] *Department of Research Development and Clinical Investigation, Children's Mercy Hospital, Kansas City, MO*

Si aucun signe de gravité

Déshydratation <5%

Diurèse conservée (>2cc/Kg/jr)

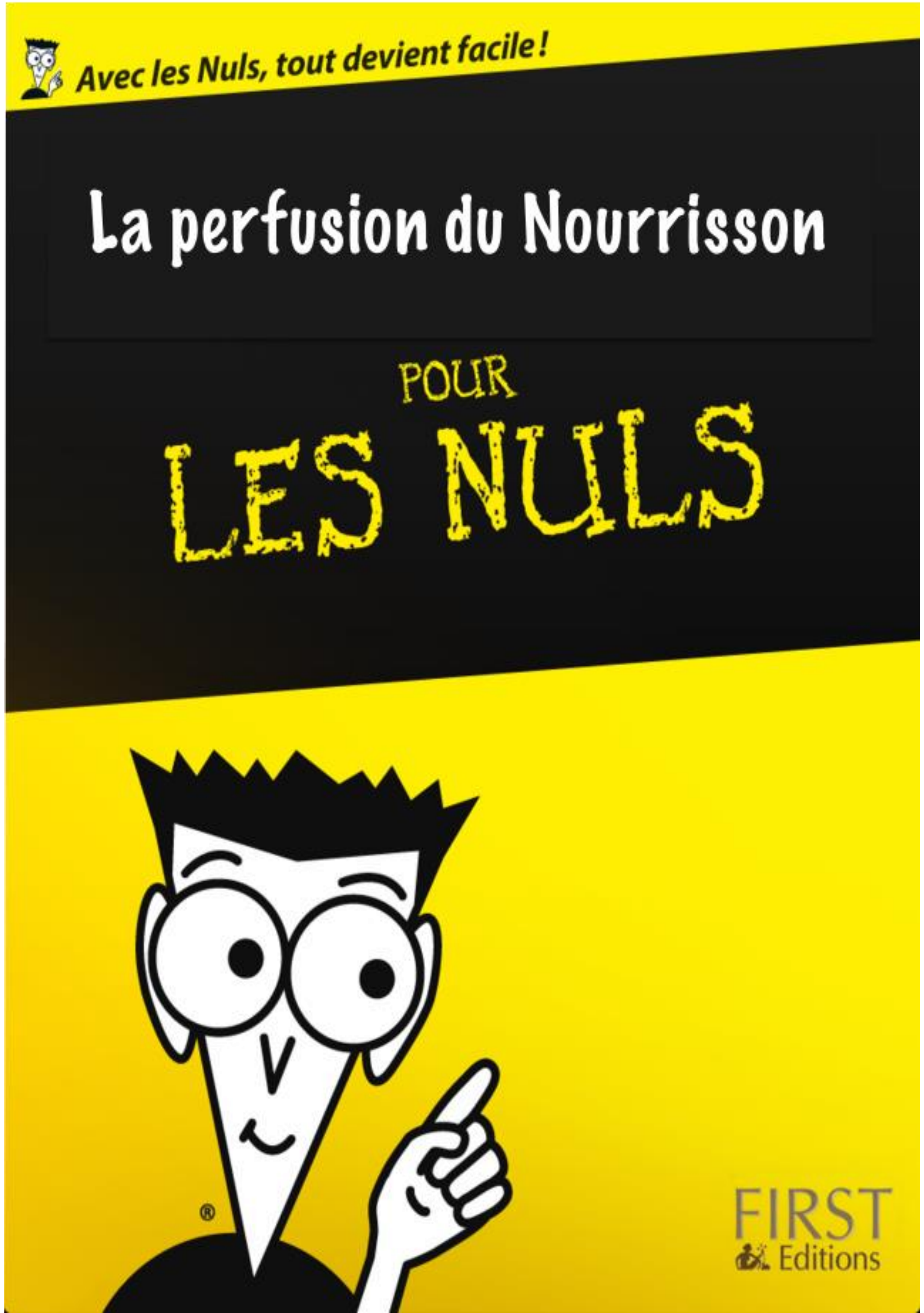
BES et GDS normaux

↓

Perfusion de maintenance

Solutés balancés et isotoniques

150mL/Kg/jour (0-3 mois)


Avec les Nuls, tout devient facile !
La perfusion du Nourrisson
POUR
LES NULS
FIRST
Editions

La perfusion du nourrisson <1 an

<u>Proscrire les solutés hypotoniques</u>

Risque d' ++ HYPONATREMIE ++

(encéphalopathie hyponatrémique, décès)

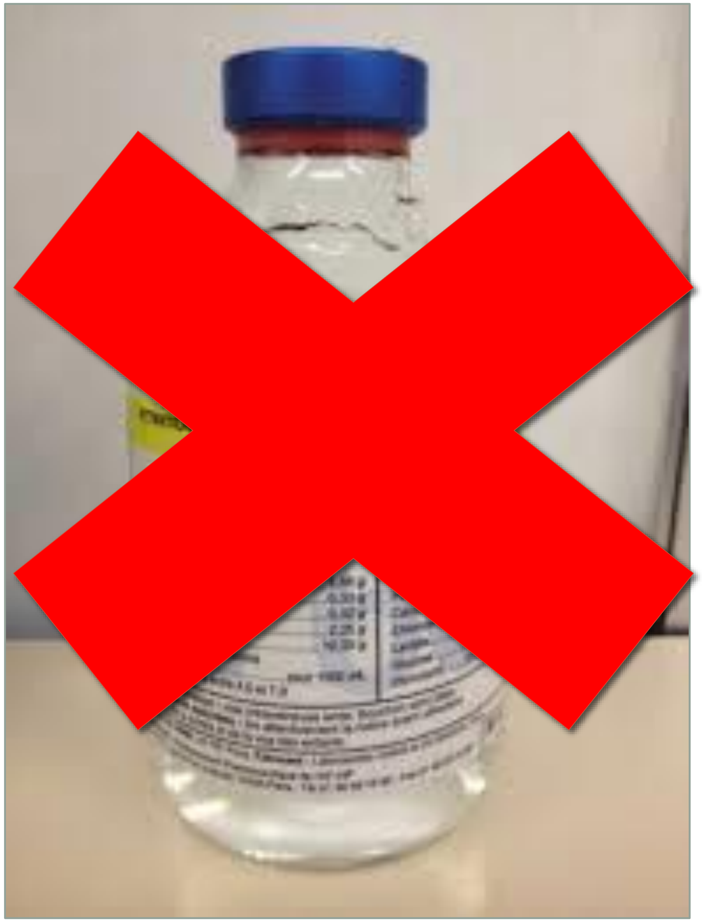
Vomissements

Stress péri-opératoire

Augmentation de la sécrétion d'ADH

La perfusion du nourrisson <1 an

La perfusion du nourrisson <1 an



La perfusion du nourrisson <1 an

La perfusion du nourrisson <1 an

- 150mL/kg/jour (0-3 mois)

  - G10% (< 1 mois)
  - ou G5% (> 1 mois)

  - + NaCl 4 mEq/Kg/jour
  - + KCl 3 mEq/Kg/jour
  - + Gluconate de Calcium 10% 2 mL/Kg/jour


HELP!

La perfusion du nourrisson <1 an

- **NaCl 10%**
  - 10 mL = 1g = 17mEq
  - 1mL = 1,7mEq
  - **1mEq=0,59mL**
  - 4mEq/kg/jr = 2,35mL/Kg/jr

- **KCl 10%**
  - 10 mL = 1gr = 13mEq
  - 1mL = 1,3mEq
  - **1mEq=0,77mL**
  - 3mEq/Kg/jr = 2,3mL/Kg/jr

La perfusion du nourrisson <1 an

Nourrisson de 3 semaines de 4Kg

G10% : 150mLx4Kg = **600 mL**

+

- **NaCl 10%** : 4mEq/kg/jr = 2,35mL x 4Kg = **9,4 mL/jr**
- **KCl 10%** : 3mEq/kg/jr = 2,3mL x 4Kg = **9,2 mL/jr**
- **Gluconate de Ca 10%** = 2mL x 4Kg = **8 mL/jr**

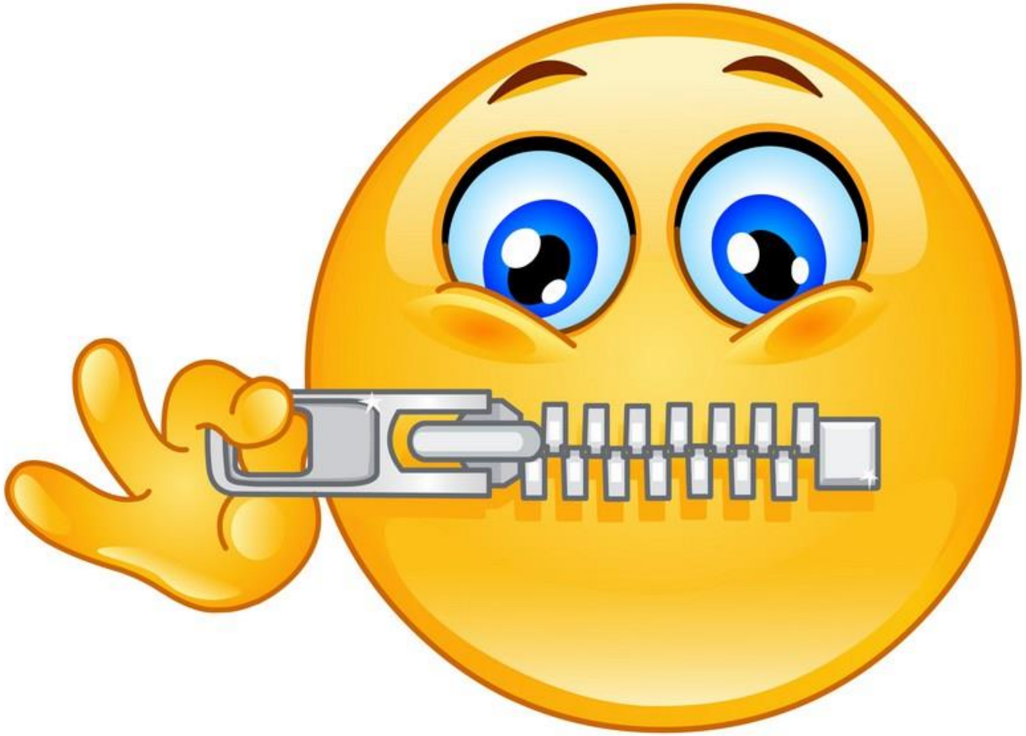
Volume total = **627 mL/ jour** soit **26 mL/heure**

La perfusion du nourrisson <1 an

- **Risque d'erreurs +++**
  - Si équipe peu habituée
  - Lors de la prescription
  - Lors de la reconstitution du soluté

- Possibilité d'utiliser solutés type Bionolyte (G5+4+2) le temps du transfert

Pylorotomie extra-muqueuse



Conclusion

- Pathologie fréquente, d’excellent pronostic
- Etiopathogénie imprécise
- Facteurs de risques multiples, environnementaux++
- Diagnostic de plus en plus précoce : Ne pas attendre les signes physiques ou biologiques pour évoquer le diagnostic
- Confirmation : échographie
- Réanimation hydro électrolytique+++
- Pylorotomie extra-muqueuse