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Detection, Evaluation, and Management of Familial Hypercholesterolemia in Children and Adolescents


December 5th, 2023



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Land Acknowledgement


I acknowledge that UBC and BC Children's
Hospital Research Institute operate on the
traditional, ancestral, and unceded territory of the
Coast Salish peoples - x^wməθk^wəyəm
(Musqueam), Skwxwú7mesh (Squamish), and
SəlNílwətaʔ/Selilwitulh (Tseil-Waututh) Nations.

Presenters






Kevin Harris, MD, MHSc
Interventional Pediatric Cardiologist
BC Children's Hospital



Venessa Thorsen, BSc (Hons)
MSc Candidate, UBC

Session Outline



- 1 Introduction, Learning Objectives, Activity Registration
- 2 Case-Based Learning (CBL): Screening, Diagnosis and Treatment of Pediatric Dyslipidemias
- 3 Contextualizing AAP and CCS Guidelines for Pediatric Dyslipidemias: 2008-Present
- 4 Helpful Resources and Closing Remarks
- 5 Q&A Period with Dr. Kevin Harris and Final Reflection Activity

Learning Objectives



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- 1 Understand the prevalence of Familial Hypercholesterolemia in Canada and the role of screening
- 2 Recall diagnostic criteria for Familial Hypercholesterolemia
- 3 Describe appropriate cholesterol screening tests for children
- 4 Formulate treatment plans for children and adolescents with Familial Hypercholesterolemia
- 5 Recall and apply guideline recommendations to clinical scenarios


Consent Form



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If you consent for data collection for a graduate thesis project, we ask that you complete a short form by scanning the QR code on screen, clicking the link provided in the chat or visiting the link below:

<https://rc.bcchr.ca/redcap/surveys/?s=HNMJHE7NA9J8NNCW>



REDCap
Research Electronic Data Capture



Demographics Survey



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Please complete the following short demographics survey

- QR code, link in chat or visit the link below:

<https://rc.bcchr.ca/redcap/surveys/?s=HE3N3443A7X4YLRA>

This survey will ask that you create a unique study ID for polling activities in the session

IMPORTANT: Please create your ID based on the street you grew up on followed by your favourite colour, in all caps

Example: CAMBIEPURPLE

This ID will be used by the study team to keep your responses anonymous, but still link your participation across platforms



PollEverywhere Registration



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This module includes interactive polling. To participate, we ask that you sign up for PollEverywhere and create a unique ID

IMPORTANT: Please create your ID based on the street you grew up on followed by your favourite colour, in all caps

Example: CAMBIEPURPLE

Please register for polling activities by scanning the QR code on screen, clicking the link provided in the chat, or visiting

<https://pollev.com/venessathorsen117>

This ID will be used by the study team to keep your responses anonymous, but still link your participation across platforms

 Poll Everywhere



Baseline Guideline Familiarity

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Poll Everywhere


Have you encountered a child with
Familial Hypercholesterolemia (FH)
in your practice before?




Please complete the first polling activity. If the activity does not automatically appear onscreen, you can follow the link provided in the chat or visit:
https://PollEv.com/clickable_images/GgeVIRA3scEtezbcJ7YAD/respond

Baseline Guideline Familiarity


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Poll Everywhere


How comfortable do you feel diagnosing/managing
primary dyslipidemias such as Familial
Hypercholesterolemia in your pediatric patients?





Please complete the polling activity. If the activity does not automatically appear onscreen, you can follow the link provided in the chat or visit:
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Baseline Guideline Familiarity


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 **Poll Everywhere**


Do you feel comfortable prescribing statins to children with primary dyslipidemias such as Familial Hypercholesterolemia (FH)?





Please complete the polling activity. If the activity does not automatically appear onscreen, you can follow the link provided in the chat or visit:
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Baseline Guideline Familiarity


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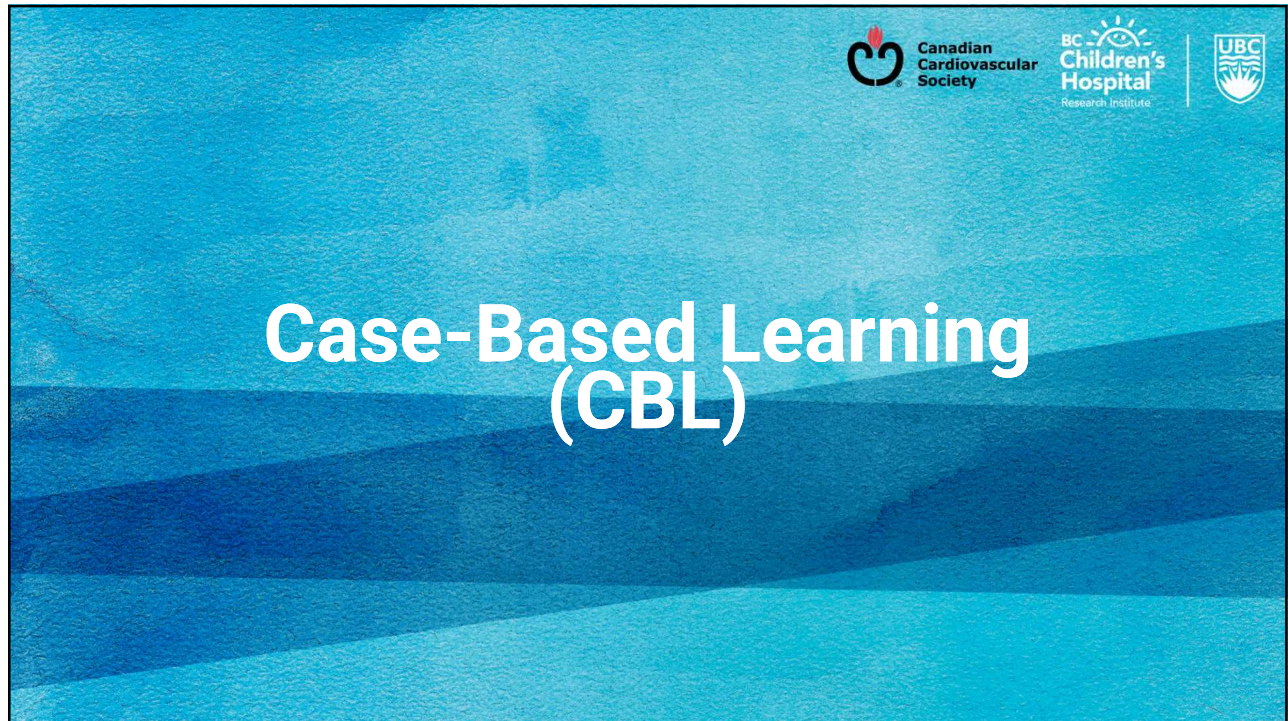


 **Poll Everywhere**

At baseline, how familiar are you with the content of the Canadian Cardiovascular Society's 2022 Clinical Practice Guidelines for Pediatric Dyslipidemias?



Please complete the polling activity. If the activity does not automatically appear onscreen, you can follow the link provided in the chat or visit:
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Case-Based Learning (CBL)



Patient Scenario #1

Dania is a healthy 10-year-old female patient in your office for developmental assessment.

You find their development to be normal.



Please complete the series of polling questions regarding your patient, Dania.

If the activity does not automatically appear onscreen, you can follow the link provided in the chat or visit:

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Screening



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Key Points: Screening



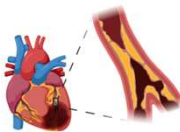
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Identifying Children with Genetic Dyslipidemias is Critical

Pediatric FH is clinically silent but easy to detect with a simple blood test. While it is clinically silent, atherosclerosis begins at birth and is progressive.



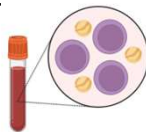
Selective Screening: Insufficient

Selective screening based on family history alone is **unreliable** and misses 30-60% of all children with dyslipidemias.



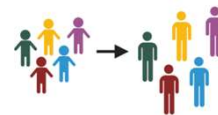
Full Lipid Profiles are the Recommended Screening Method

Fasting or non-fasting full lipid profiles are the recommended screening method to detect **abnormally high non-HDL or LDL-C**.



Universal Pediatric Lipid Screening

The Canadian Cardiovascular Society recommends **universal lipid screening** for children between the ages of 2-10.





Patient Scenario #2

Results have returned from the fasting lipid profile you ordered for your 11-year-old patient Mimi.

She has an LDL-C level of 5.2 mmol/L (Normal range <2.8 mmol/L). This result is consistent with a previous test ordered 3 months ago despite dietary and lifestyle changes.

She does not have any other systemic disease but has a positive family history of Familial Hypercholesterolemia.


 **Poll Everywhere**

Please complete the series of polling questions regarding your patient, Mimi.



If the activity does not automatically appear onscreen, you can follow the link provided in the chat or visit:
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Diagnosis



Key Points: Diagnosis

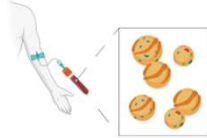


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Fasting Lipid Profile Markers of FH



An average LDL cholesterol level of **>4.1 mmol/L or >3.4 mmol/L with a family history** of elevated LDL cholesterol, premature CAD and/or genetic diagnosis is sufficient to diagnose FH in pediatric patients.

Two fasting lipid profiles obtained 2 weeks to 3 months apart should be obtained to confirm the diagnosis.

Physical Exam Findings May Aid Diagnosis

Physical examination findings including xanthelasmas, tendon xanthomas (A), eruptive xanthomas over extensor surfaces and buttocks (B), Lipemia retinalis (C) and corneal arcus (D) are suggestive of an undiagnosed dyslipidemia.



****Physical findings are rare in young children and should not preclude screening****

Genetic Testing

When accessible, genetic testing is useful to achieve definitive diagnoses of FH and other genetic dyslipidemias.



Patient Scenario #3

You have diagnosed Easton, a 9-year-old patient at your office, with Familial Hypercholesterolemia (FH) based on repeated lipid profiles and a positive family history.

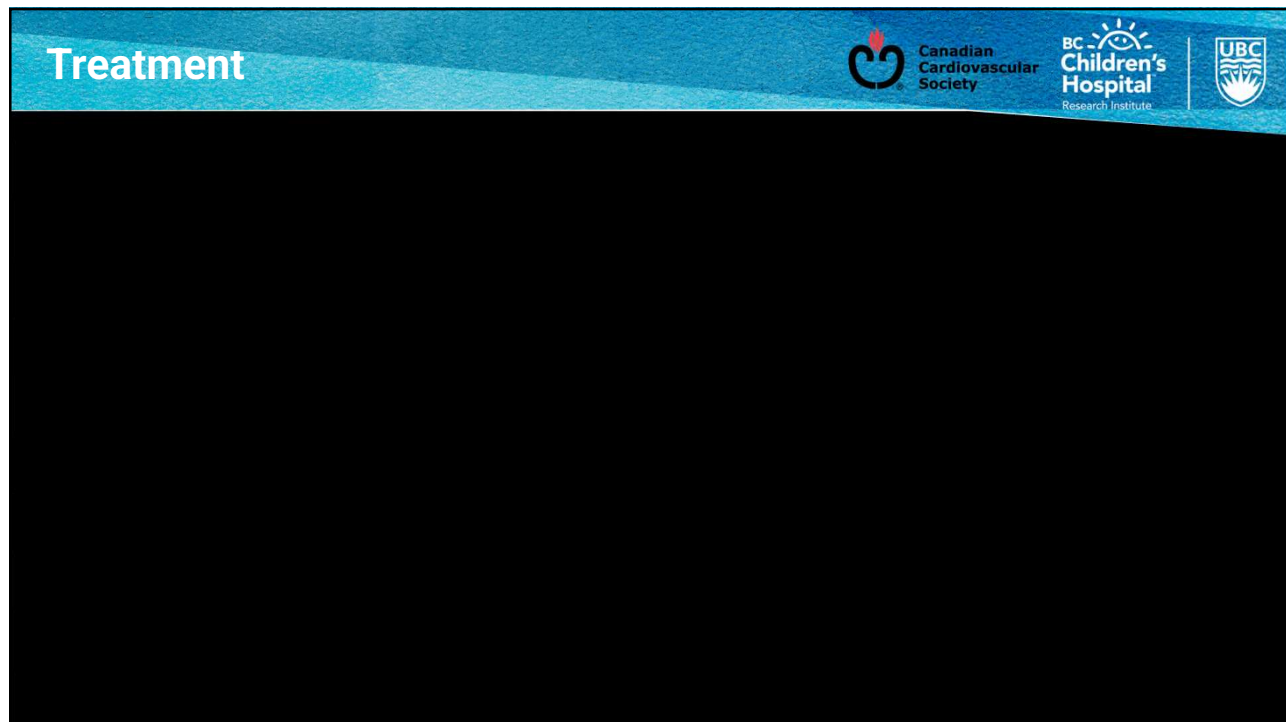
 **Poll Everywhere**

Please complete the series of polling questions regarding your patient, Easton.







If the activity does not automatically appear onscreen, you can follow the link provided in the chat or visit:

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Key Points: Treatment




Statins for Children with FH

- **First line** medical therapy for Familial Hypercholesterolemia and other pediatric dyslipidemias.
- Monitor for comorbidities over the course of treatment.



Statin Therapy is Safe and Effective in Children

- Safe and effective for patients 8+ when LDL-C remains above treatment thresholds.
- **Well-tolerated** in pediatric patients.
 - **No effect on growth and development.**
- Muscle-related symptoms very rare in children.
 - No cases of pediatric rhabdomyolysis documented.




Lifestyle Counselling

- **Diet and exercise are important but not sufficient to treat genetic dyslipidemias such as Familial Hypercholesterolemia alone.**



To help establish lifestyle counselling and recommendations, consider referring to:

- A pediatric lipid specialist
 - Dietician
- Preventive Cardiology Services at BC Children's Hospital.

Contextualizing the New Guidelines

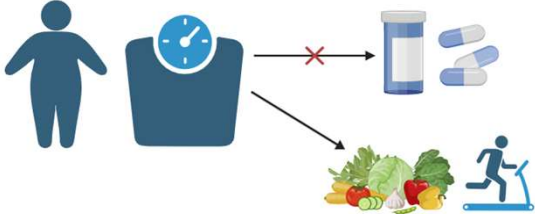


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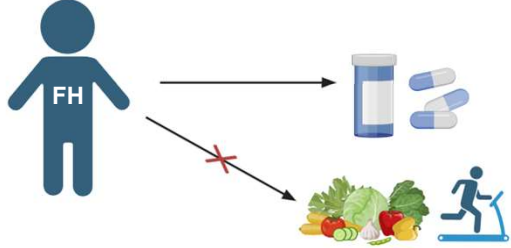
AAP Guidelines (2008)

- Released as issues of childhood obesity were coming to light, developed for this population
- Recommendations include:
 - Selectively screening for patients with positive family history of CVD and/or obesity
 - Limiting dietary intake of saturated and trans fats, cholesterol and carbohydrates
 - Increasing physical activity level
- Retired in 2012**




AAP Guidelines (2020)



- The AAP now recommends universal screening of children ages 9-11 years for dyslipidemias
- Statins safe and effective for children
- Diet and exercise important but insufficient on their own for genetic dyslipidemias
- Aligned with 2022 CCS and CPS recommendations**




Primary vs. Secondary Dyslipidemias



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Primary Dyslipidemias



- FH is most common, other less common dyslipidemias may also be detected (see CCS guidelines)
 - FH is a heritable deficiency in LDL-C receptor causes 2-3x higher blood cholesterol
 - 1 in ~250 Canadians
- Not sufficiently managed by diet and exercise alone


↓

First line Therapy: Statins

Lipid-lowering medications like statins are essential to manage primary dyslipidemias

Early involvement of dietician and 60mins/day MVPA

Secondary Dyslipidemias



- Acquired condition
 - Secondary to **obesity**, diabetes, renal and liver disease, and/or medications
- Increasing prevalence
- Sufficiently managed by diet and exercise in most cases

↓

First line Therapy: Diet and Exercise

Diet


- Early involvement of dietician
- CHILD-1 diet (<30% calories from fat, 8-10% of daily caloric intake)
- CHILD-2 diet (<7% saturated fat and <10% monounsaturated fat)

Exercise


- 60mins/day MVPA

If statins are started, diet and exercise continue to be important

AAP Guidelines vs. CCS Guidelines



American Academy of Pediatrics
DEDICATED TO THE HEALTH OF ALL CHILDREN™



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	Acceptable	Borderline	Abnormal	Acceptable	Borderline	Abnormal
Total cholesterol (TC)	< 4.4mmol/L	4.4 - < 5.1mmol/L	≥ 5.2mmol/L	< 4mmol/L	4.4 - < 5.2mmol/L	≥ 5.2mmol/L
LDL-C	< 2.8mmol/L	2.8 - < 3.3mmol/L	≥ 3.4 mmol/L <small>*plus positive family history</small>	< 2.8mmol/L	2.8 - < 3.4mmol/L	≥ 3.4 mmol/L <small>*plus positive family history</small>
HDL-C	> 1.2mmol/L	1.0 - 1.2mmol/L	< 1.0mmol/L	> 1.2mmol/L	1.0 - 1.2mmol/L	< 1.0mmol/L
Triglycerides						
0-9 years	< 0.85mmol/L	0.85 - <1.12mmol/L	≥ 1.13mmol/L	< 0.8mmol/L	0.8 - <1.1mmol/L	≥ 1.1mmol/L
10-19 years	< 1.02mmol/L	1.02- <1.46mmol/L	≥ 1.47mmol/L	< 1.0mmol/L	1.0- <1.5mmol/L	≥ 1.5mmol/L

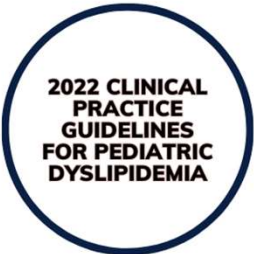
Closing

Resources

Guidelines: Update for code on site <https://online.ubc.ca/clinical-practice-guidelines-for-pediatric-dyslipidemia>



Session K presented complete

Flow Chart pediatric complete



2022 CLINICAL PRACTICE GUIDELINES FOR PEDIATRIC DYSLIPIDEMIA

Abnormal:
TC ≥ 5.2mmol/L
LDL-C ≥ 4.1mmol/L or ≥ 3.5 and positive family history
Non-HDL-C ≥ 3.75mmol/L
Triglycerides ≥ (0-9 years) 1.1mmol/L
Triglycerides ≥ (10-19 years) 1.5mmol/L
HDL-C < 1.0mmol/L

Universal screening at ages 9-11 and 17-21

Selective screening in children with a FHx of premature CVD and/or Hx of risk factors (obesity, hypertension, DM Type II)

Cascade screening for 1st degree relatives of those with FH or other dyslipidemias

Order fasting or non-fasting lipid profile for universal and selective screening

Offer cascade screening for 1st degree relatives of patients with primary dyslipidemias

2 abnormal fasting lipid profiles within 2 weeks-3 months of each other and/or positive genetic testing results for FH or other dyslipidemias

Initiate statin therapy if target lipid levels are not reached despite previous interventions

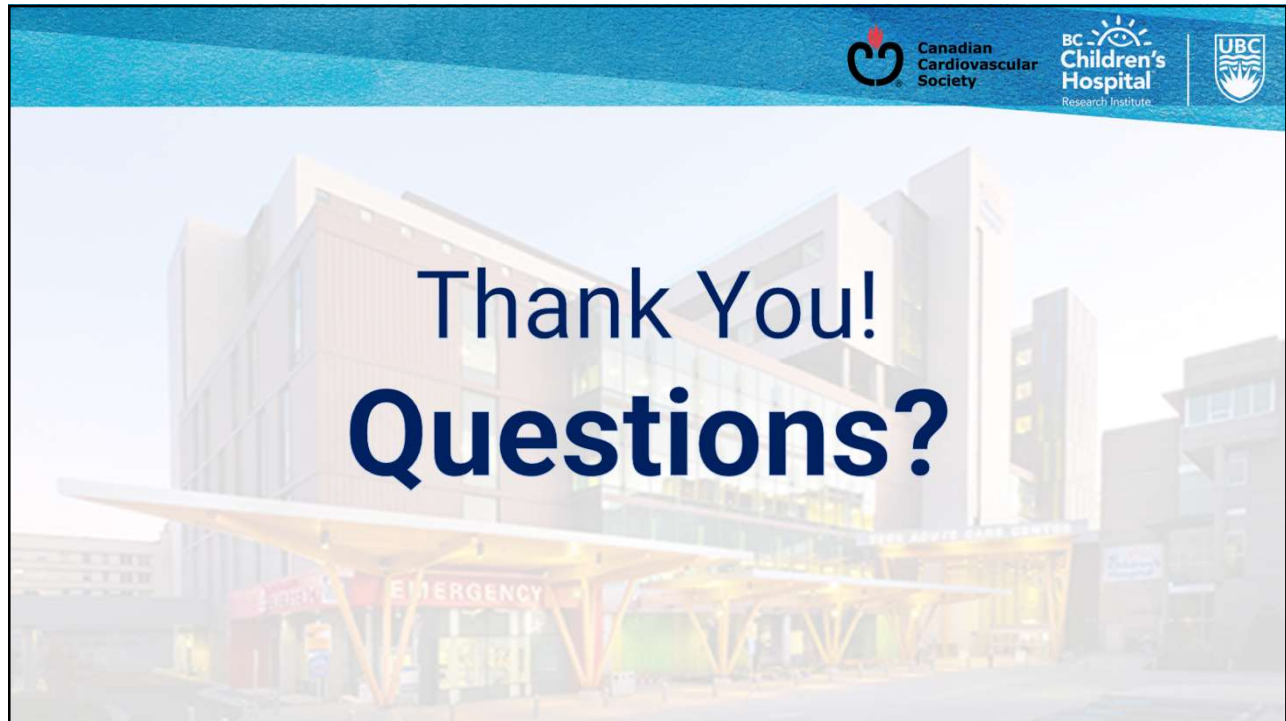
Implement dietary/lifestyle modifications

Consider referral to pediatric lipid specialist

After starting statin therapy, routine monitoring for:
a) safety (monitor kidney/liver function with blood test)
b) LDL-C treatment targets

REB Ethics # H23-00439



V1.0 (July 16, 2023)

A slide with a blue header containing logos for the Canadian Cardiovascular Society, BC Children's Hospital Research Institute, and UBC. The main content area has a white background with the following text:

You will be contacted with a short post-session survey in one month, which will help us to improve this course and optimize care for children with dyslipidemias in BC.

We ask that you PLEASE complete this survey.

For study related inquiries, please do not hesitate to contact Venessa Thorsen at cardioresearch@bcchr.ca.



Before you go...



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Please complete the final reflection.
We anticipate this will take no more
than 3-5 minutes.



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