Case presentation

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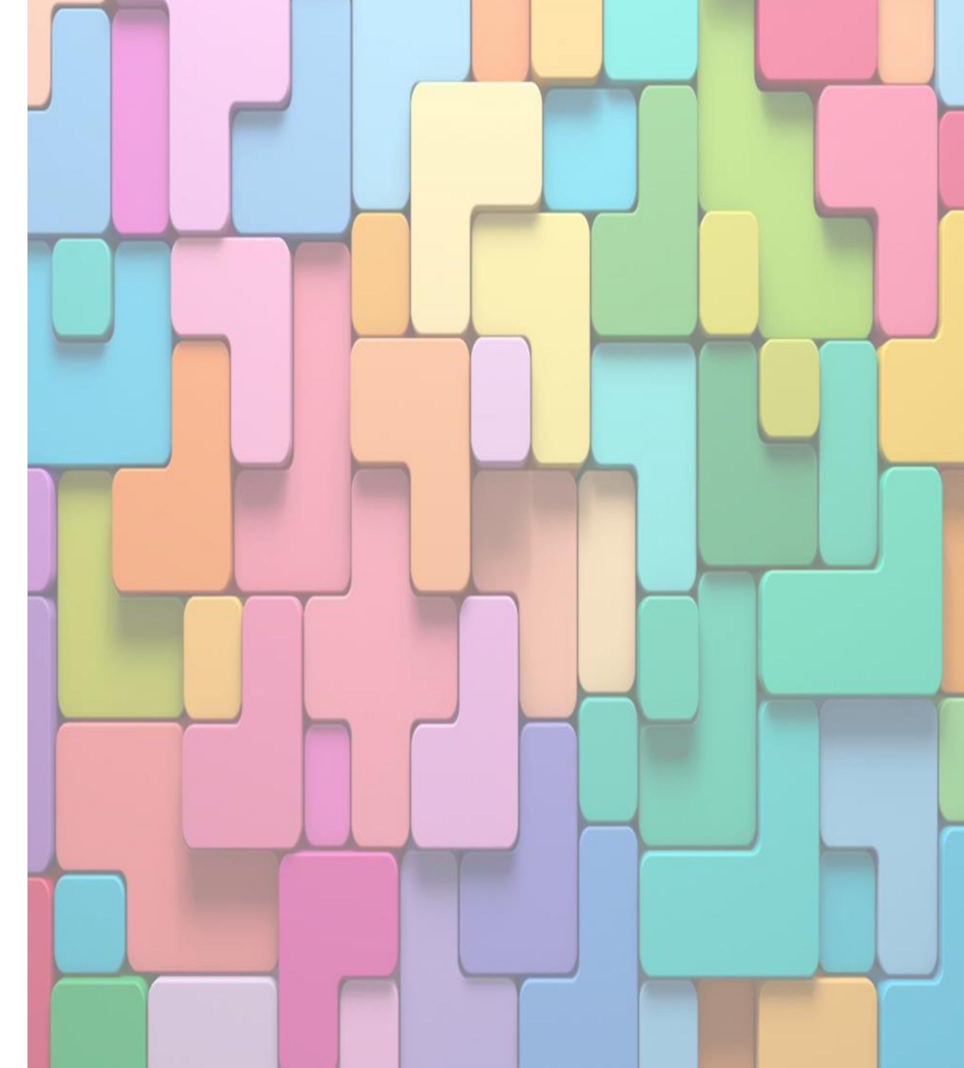


Objective

Case Overview

Going back …

Recommendation …

Case Overview

Master M

9-year-old boy :

Presenting complaint

- Concern about “Inattention”
- Behavioural and learning concern

Background

<u>Background</u>

1. Autism Spectrum Disorder (2024)
2. ADHD
3. Anxiety
4. Dysgraphia (diagnosis made on formal testing with RTLB)
5. Complex cardiac diagnoses

Complex cardiac diagnosis

1. Criss-cross concordant atrioventricular cardiac connections with double outlet right ventricle (anterior and leftward ventricle with aorta leftward and anterior with pulmonary atresia)

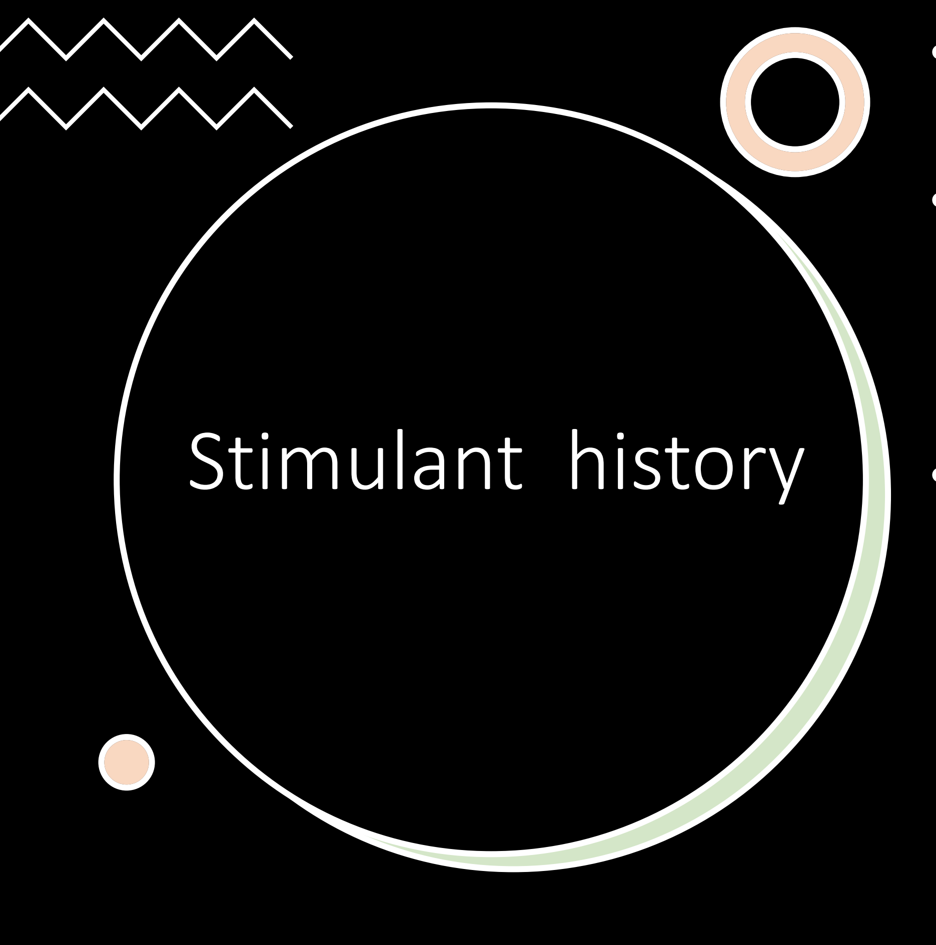
2.PDA ligation, Bidirectional Glenn with atrial septectomy, left pulmonary artery balloon dilatation, coiling of the right internal mammary artery collateral, cardiac Fontan with Gortex graft, left pulmonary artery plasty with bovine pericardium

3. Changed from Warfarin to Aspirin for Fontan thromboprophylaxis January 2020

Aspirin 100mg daily
Fluoxetine 20mg daily

- Learning concern. The school is struggling to get him to do any work
- Refuse to do if “unable to do it” or “not want to do it”. If forced, he will become angry
- At home, difficulty to follow through with instructions, and ask mum to stay with him to stay on task
- The teacher is managing things with a specialised learning plan but unable to progress with his learning.
- Very impulsive and gets distracted easily.

- Upset If there is a reliever teacher (routine change)
- He does not like things to be rushed and switching tasks might be a bit difficult. He can be inflexible.
- Some sensory issues. Loud noises
- No obvious bullying or stressors.
- Most settled at school at this time. He has a few friends.
- Better on fluoxetine



Stimulant history

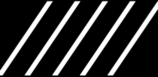
- Previously on Rubifen 10mg morning and 5mg at noontime
- Stopped (mid 2023) because of lots of tummy pain and jittery. He had some pain prior to starting medication but pain got worse. Not help.
- At that time, did not like with school and was very anxious. When the medication stopped, there was some improvement with the pain, but things got better when there were changes made with his behavioural/ learning plans at school.

Cardiac

- Recent review and Echocardiogram
- Stable from cardiac perspective
- Continue Fontan surveillance
- Ongoing follow up

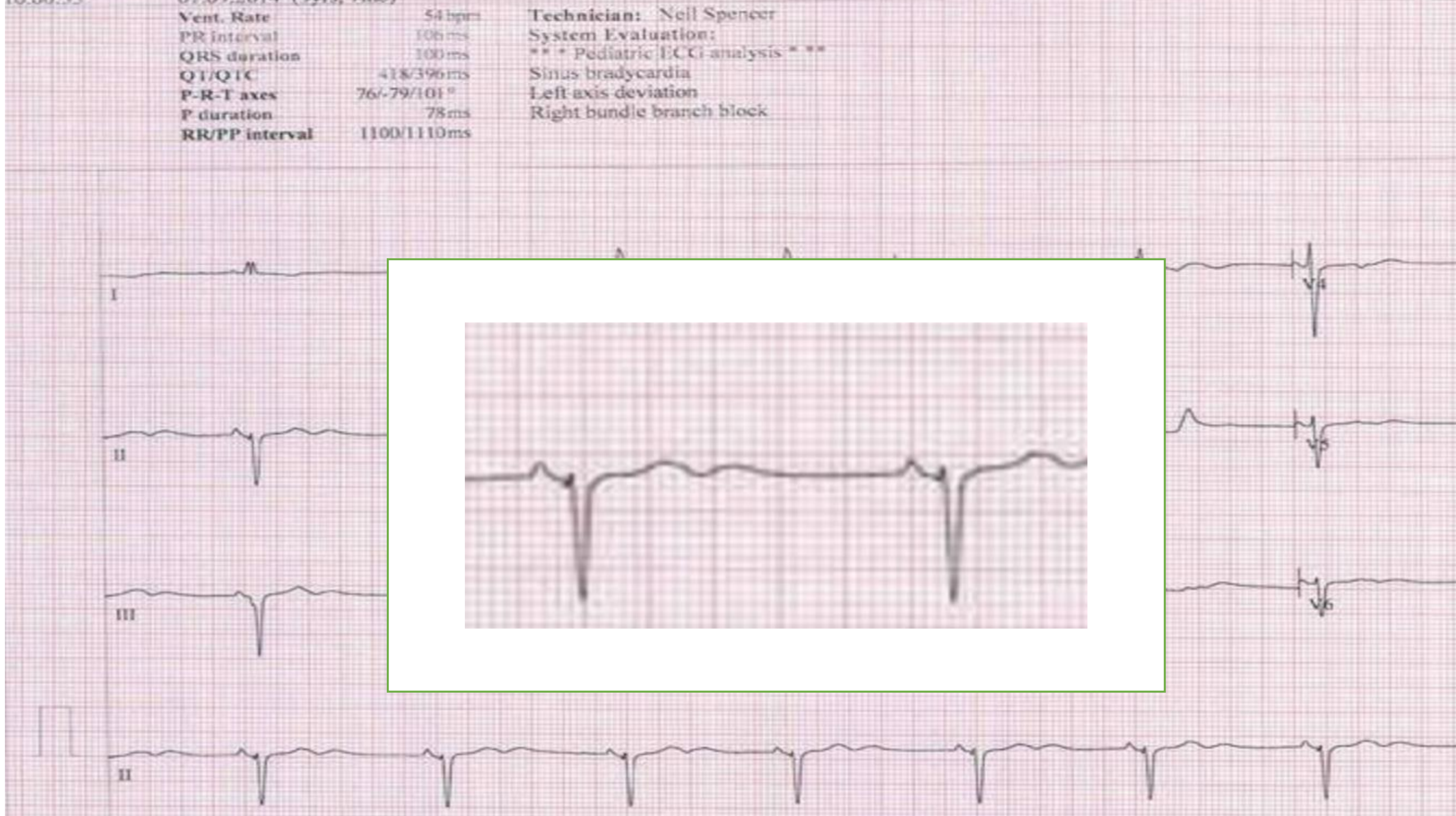
Clinical examination

- Not seen yet…
- Previous :
- Blood pressure was 110/60. Oxygen saturations 98% on room air.
- Weight 28.4kg (9-25th centile)
- Height 128.4cm (9-25th centile)
- His heart sounds were normal with no murmurs detected. Respiratory and abdomen examination was unremarkable



Impression/Plan

- Reconsider stimulant medication.
- Get a height, weight, and blood pressure before starting this medication.
- Look at his ECG (may need a repeat ECG)



Vent. Rate 54 bpm
PR interval 106 ms
QRS duration 100 ms
QT/QTC 418/396 ms
P-R-T axes 76/-79/101 °
P duration 78 ms
RR/PP interval 1100/1110 ms
Technician: Neil Spencer
System Evaluation:
** * Pediatric ECG analysis * **
Sinus bradycardia
Left axis deviation
Right bundle branch block
I
II
III
II
V4
V5
V6

Going back …

Marcrolid in Cystic fibrosis (CF) and non-cystic fibrosis(CF) bronchiectasis

Long-term effects of azithromycin in patients with cystic fibrosis

Clémentine Samson [a, b], Aline Tamalet [a], Hoang Vu Thien [c], Jessica Taytard [a, f], Caroline Perisson [a], Nadia Nathan [a], Annick Clement [a, d], Pierre-Yves Boelle [d, e], Harriet Corvol [a, d, f, *]

Cystic Fibrosis Pulmonary Guidelines

Chronic Medications for Maintenance of Lung Health

Peter J. Mogayzel, Jr.[1], Edward T. Naureckas[2], Karen A. Robinson[3], Gary Mueller[4], Denis Hadjiliadis[5], Jeffrey B. Hoag[6], Lisa Lubsch[7], Leslie Hazle[8], Kathy Sabadosa[8], Bruce Marshall[8], and the Pulmonary Clinical Practice Guidelines Committee*

Long-term azithromycin for Indigenous children with non-cystic-fibrosis bronchiectasis or chronic suppurative lung disease (Bronchiectasis Intervention Study): a multicentre, double-blind, randomised controlled trial

Patricia C Valery, Peter S Morris, Catherine A Byrnes, Keith Grimwood, Paul J Torzillo, Paul A Bauert, I Brent Masters, Abbey Diaz, Gabrielle B McCallum, Charmaine Mobberley, Irene Tjhung, Kim M Hare, Robert S Ware, Anne B Chang

Summary

Systematic Review and Meta-Analysis

Medicine

OPEN

Meta-analysis of macrolide maintenance therapy for prevention of disease exacerbations in patients with noncystic fibrosis bronchiectasis

Donghai Wang, MS[a,b], Wenlong Fu, MS[a], Jihong Dai, MD[a,b,*]

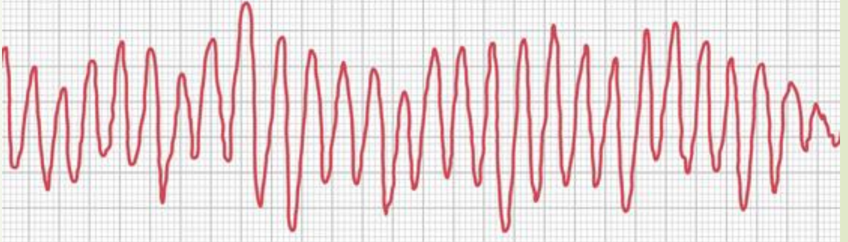
Macrolide

High plasma concentration

Anti-inflammatory properties.

Long half-life

Prolong QT



Torsades de pointes (TdP) can lead to symptoms of:

- Dizziness and syncope.
- Asymptomatic
- First presentation can be sudden unexpected death

Congenital Long QT syndrome (LQTS)

- Inherited genetic disorder
- Mutation of the heart ion channels
- Incidence 1 in 2000.
- 2000 to 3000 sudden unexpected deaths yearly in America.

Macrolide related QT prolongation

Macrolide is classified Known Risk(KR)

Risk increase

- Elderly
- Female gender
- Underlying cardiac illness
- Concomitant use of other drugs
- Acutely unwell patients
- Electrolyte abnormality (hypokalaemia)

Macrolide related QT prolongation

LETTER

Lack of effect of azithromycin on QT interval in children: a cohort study

Macrolides are a group of antimicrobial drugs used widely, being well known for their adverse cardiac effects. Erythromycin and clarithromycin are most commonly

Journal of Cystic Fibrosis 15 (2016) 192–195

Short Communication

An evaluation strategy for potential QTc prolongation with chronic azithromycin therapy in cystic fibrosis☆

Patrick John Lenehan [a,*], Craig M. Schramm [b,c], Melanie Sue Collins [b,c]

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[b] Pulmonary Department, Connecticut Children's Medical Center, 282 Washington Street, Hartford, CT 06106, USA
[c] Department of Pediatrics, University of Connecticut School of Medicine, 263 Farmington Avenue Farmington, CT 06030, USA

Audit on Long QT in Cystic fibrosis (CF) and non-cystic fibrosis(CF) bronchiectasis

$$QTc = \frac{QT}{\sqrt{R-R}}$$

- Audit from 2017 to 2019.
- Review 269 patient – all ECG

N=54 Cystic fibrosis

N=215 non-cystic fibrosis bronchiectasis

- On long term macrolide.

9% N=5 CF group

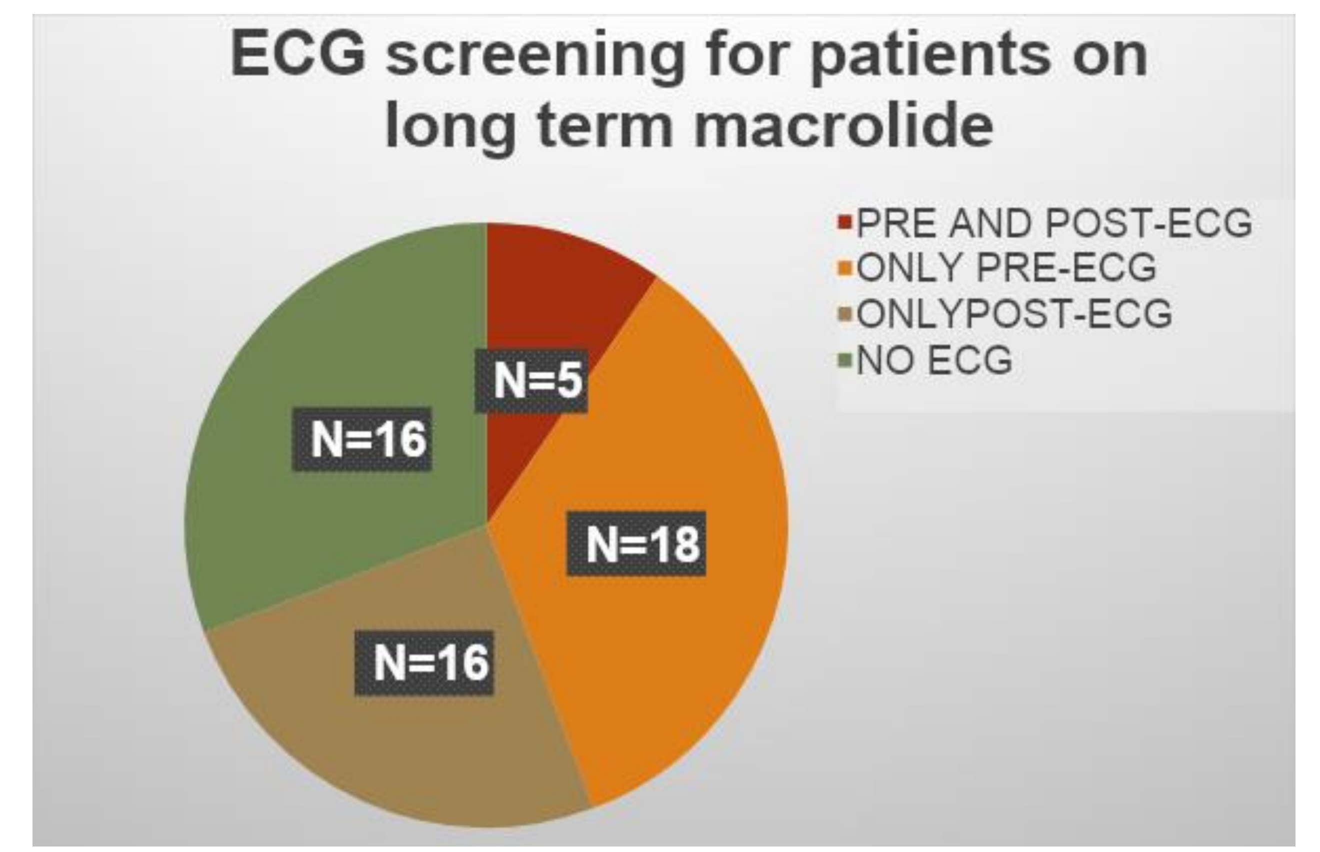
18% N=40 non-CF bronchiectasis

- Calculate QTc using Bazett formula

Concomitant medications

Concomitant medications

- CF n=4, 80%
- non-CF bronchiectasis n=41, 52.5%


ECG screening for patients on long term macrolide
PRE AND POST-ECG
ONLY PRE-ECG
ONLYPOST-ECG
NO ECG
N=5
N=18
N=16
N=16

How good are we at measuring QTc?

Found one patient with long QTc interval on a pre-macrolide ECG with no follow up ECG documented.

Difficulty to confidently work out the QTc interval of 2 patients.

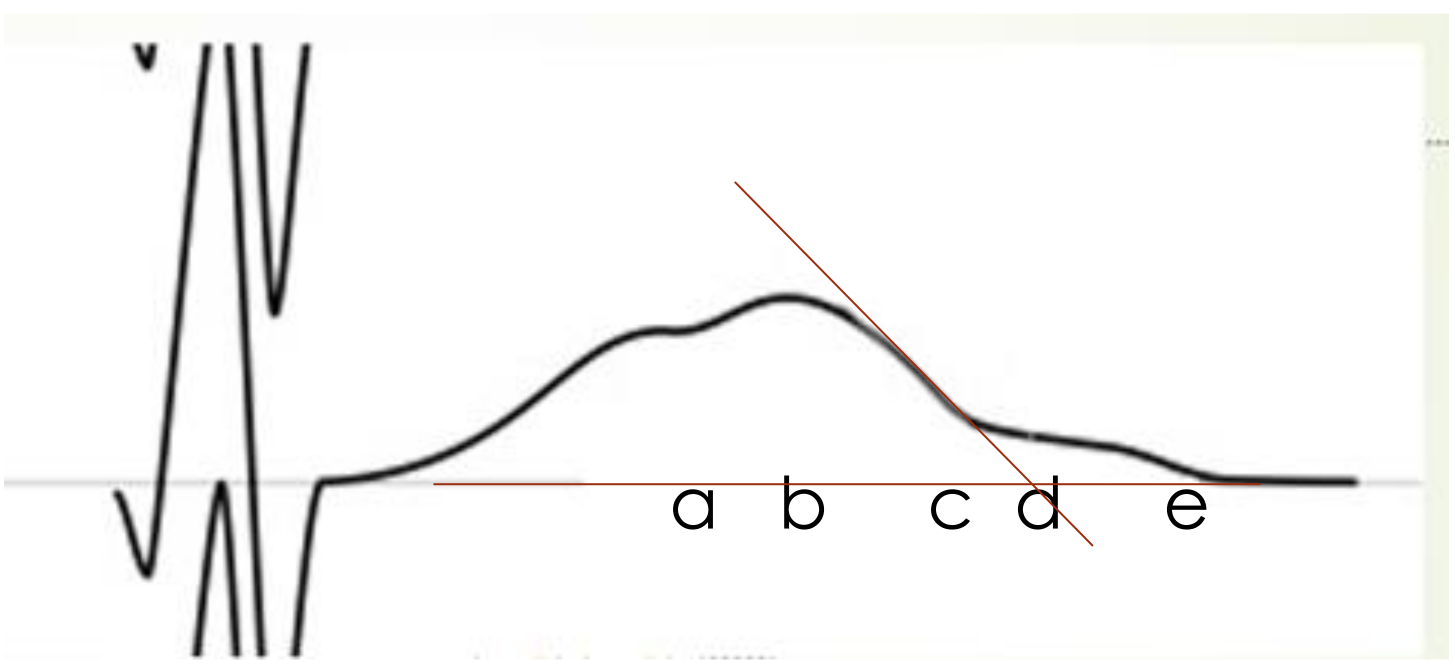
Current … Pre-treatment surveillance

An ECG before starting Azithromycin is mandatory to exclude prolongation of QTc interval.

Review current/other ongoing medicines to ensure there is no risk of interaction which will further risk QTc prolongation.

https://starship.org.nz/guidelines/bronchiectasis-management/

How good are we at measuring the QT interval?



How to measure the QT interval on an ECG…

Acronym "PQRSTU"

P - **P**referred on lead II of the ECG. Alternative, lead V5, V6 or I can be used

Q -**Q**T interval (start of Q wave to end of T wave)

R - **R**esting heart **R**ate, **R**esolution high, **R**epeat

S - **S**table **S**inus rhythm (3 consecutive)

T - **T**-wave morphology, **T**angential method[1]

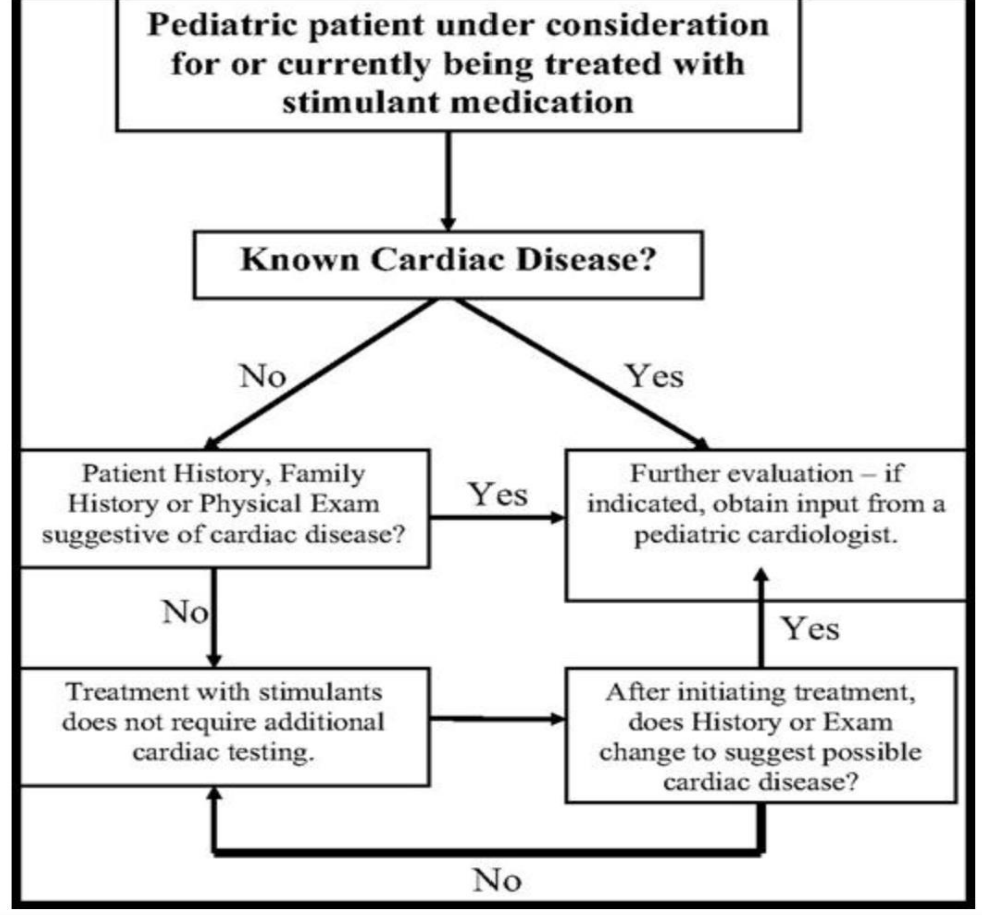
U- **U**-wave may be included. [2]
Underlying cause for long QT needs to be fix.

1. If bifid T-wave tangential method should be used on the second peak if the amplitude is > 50% of the first.Be familiar with T wave morphology for LQTS

2. Include the U-wave if the U-wave amplitude >50% of the T wave and merging into the T wave.

Recommendation

Is there recommendation for stimulant medication…



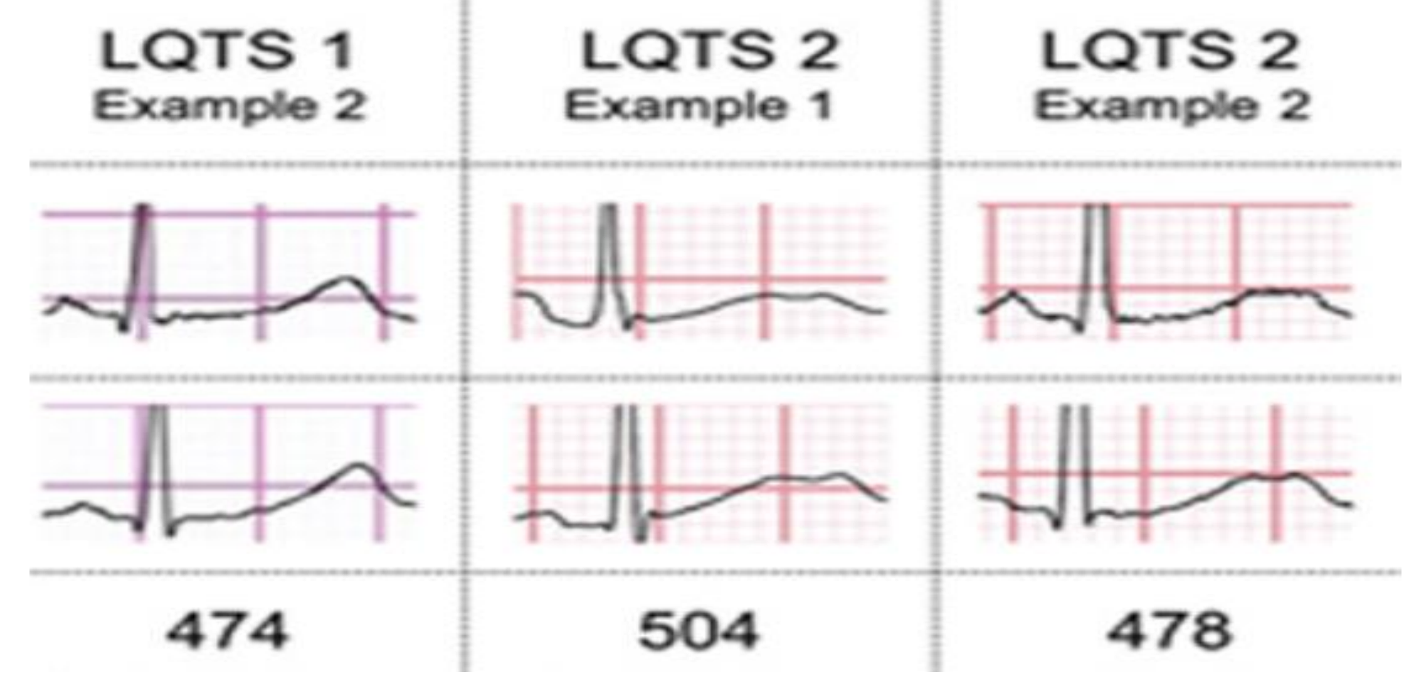
James M. Perrin, Richard A. Friedman, Timothy K. Knilans, the Black Box Working Group, the Section on Cardiology and Cardiac Surgery; Cardiovascular Monitoring and Stimulant Drugs for Attention-Deficit/Hyperactivity Disorder. *Pediatrics* August 2008; 122 (2): 451–453. 10.1542/peds.2008-1573

Take home message

ECG screening for stimulant

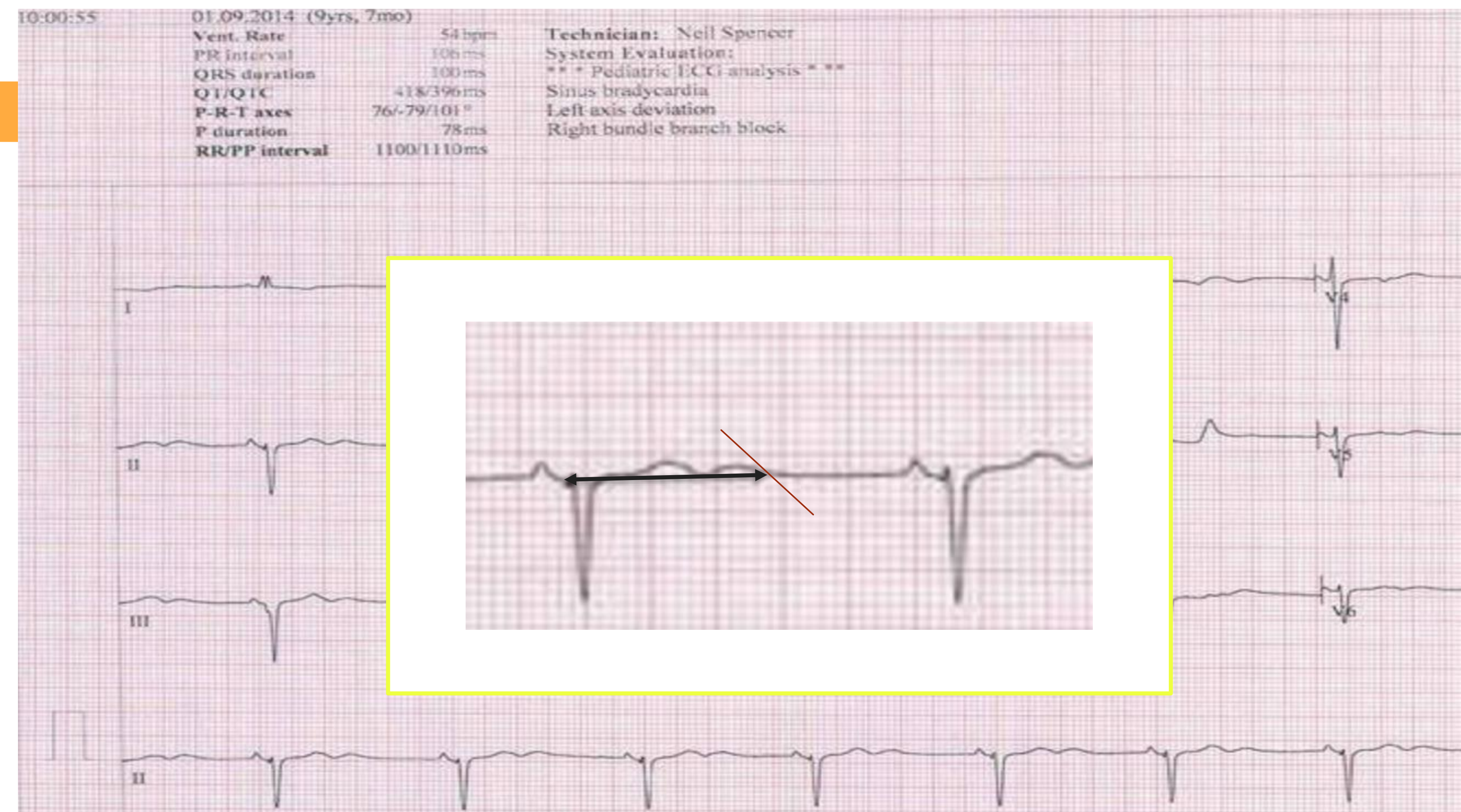
- Consider ECG screening if there is a family history of sudden unexpected death, cardiac issues or risk factor.
- Post ECG screening can be done in the next clinic visit or earlier if the patient's circumstances changes.
- Use the "PQRSTU" acronym
- Take note of the T wave morphology
- Seek a second opinion from the Cardiologist/Specialist if there is any concern/uncertain
- Be aware of drugs that can prolong QT

https://www.crediblemeds.org/



QTc

"T-Wave Morphology Analysis in Congenital Long QT Syndrome Discriminates Patients From Healthy Individuals": https://www.sciencedirect.com/science/article/pii/S2405500X16304066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System Evaluation:
*** Pediatric ECG analysis ***
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ECG…feedback on master M

- T-wave appears long, but the morphology is difficult to interpret.
- Not safe to start stimulant medication until this is clarified.
- Repeat ECG (could increase the gain on the ECG if the T wave is still difficult to measure).
- Already on a QT prolonging agent - good to be cautious about this

Thanks

Reference

- https://starship.org.nz/guidelines/bronchiectasis-management/#:~:text=Azithromycin%20in%20bronchiectasis&text=Ministry%20of%20Health%20provides%20up,treatment%20may%20be%20clinically%20indicated
- https://media.starship.org.nz/paediatric-ecg-interpretation/Paediatric_ECG.pdf
- James M. Perrin, Richard A. Friedman, Timothy K. Knilans, the Black Box Working Group, the Section on Cardiology and Cardiac Surgery; Cardiovascular Monitoring and Stimulant Drugs for Attention-Deficit/Hyperactivity Disorder. *Pediatrics* August 2008; 122 (2): 451–453. 10.1542/peds.2008-1573
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- "T-Wave Morphology Analysis in Congenital Long QT Syndrome Discriminates Patients From Healthy Individuals": https://www.sciencedirect.com/science/article/pii/S2405500X16304066