

## SUPPLEMENTAL MATERIAL

### Supplemental Tables

Voting questions Case 1:	
Q1: Case 1, this patient (asymptomatic teen, gene (-) LQTS, QTc 480 ms, athlete without symptoms, assuming that after a period of deconditioning and additional testing you are convinced he has LQTS) should be treated with a beta blocker	<ol style="list-style-type: none"> <li>1. Yes unless there is a contraindication such as asthma</li> <li>2. No</li> </ol>
Q2: Case 1, this patient (asymptomatic teen, gene (-) LQTS, QTc 480 ms, athlete without symptoms, assuming that after a period of deconditioning and additional testing you are convinced he has LQTS), should be treated with more advanced therapy if he cannot take a beta blocker	<ol style="list-style-type: none"> <li>1. Yes, LCSD</li> <li>2. Yes, ICD</li> <li>3. Yes, both</li> <li>4. No</li> </ol>
Q3: Case 1, this patient (asymptomatic teen, gene (-) LQTS, QTc 480 ms, athlete without symptoms, assuming that after a period of deconditioning and additional testing you are convinced he has LQTS) should be restricted from:	<ol style="list-style-type: none"> <li>1. All sports</li> <li>2. Competitive sports</li> <li>3. Competitive swimming, diving only</li> <li>4. No sports restrictions</li> </ol>
Q4: Case 1, Considering this patient (asymptomatic teen, gene (-) LQTS, QTc 480 ms, athlete without symptoms, assuming that after a period of deconditioning and additional testing you are convinced he has LQTS), there should be repeat commercial genetic testing on the proband or the patient if it has been > 10 years since the proband was tested or testing was done in a research lab.	<ol style="list-style-type: none"> <li>1. Yes</li> <li>2. No</li> </ol>

**TABLE I**

Last Name	First Name	QUESTION 1	QUESTION 2	QUESTION 3	QUESTION 4
Zareba	Wojciech	Yes, unless there is an contraindication such as asthma	No	Competitive sports	Yes
Wilde	Arthur	Yes, unless there is an contraindication such as asthma	No	No sports restrictions	Yes
Towbin	Jeffery	Yes, unless there is an contraindication such as asthma	Yes, LCSD	Competitive swimming, diving only	Yes
Sy	Raymond	Yes, unless there is an contraindication such as asthma	Yes, LCSD	Competitive sports	Yes
Shoemaker	Benjamin	Yes, unless there is an contraindication such as asthma	No	Competitive sports	Yes
Shimizu	Wataru	No	No	Competitive sports	No
Schwartz	Peter	Yes, unless there is an contraindication such as asthma	Yes, LCSD	Competitive sports	Yes
Schulze-Bahr	Eric	Yes, unless there is an contraindication such as asthma	No	Competitive sports	Yes
Roden	Dan	Yes, unless there is an contraindication such as asthma	No	Competitive swimming, diving only	Yes
Roberts	Jason	Yes, unless there is an contraindication such as asthma	No	No sports restrictions	Yes
Priori	Silvia	Yes, unless there is an contraindication such as asthma	No	Competitive sports	Yes
Perez	Marco	Yes, unless there is an contraindication such as asthma	No	Competitive sports	Yes
Lubitz	Steven	Yes, unless there is an contraindication such as asthma	No	Competitive swimming, diving only	Yes

Krahn	Andrew	Yes, unless there is a contraindication such as asthma	No	No sports restrictions	Yes
Kaufman	Elizabeth	Yes, unless there is a contraindication such as asthma	Yes, LCSD	Competitive swimming, diving only	Yes
Etheridge	Susan	Yes, unless there is a contraindication such as asthma	Yes, LCSD	Competitive swimming, diving only	Yes
Eckhardt	Lee	Yes, unless there is a contraindication such as asthma	Yes, LCSD	No sports restrictions	Yes
Culter	Michael	Yes, unless there is a contraindication such as asthma	No	No sports restrictions	Yes
Chung	Mina	Yes, unless there is a contraindication such as asthma	No	No sports restrictions	Yes
Cerrone	Marina	Yes, unless there is a contraindication such as asthma	No	No sports restrictions	Yes
Behr	Elijah	Yes, unless there is a contraindication such as asthma	No	No sports restrictions	Yes
Aziz	Peter	Yes, unless there is a contraindication such as asthma	No	No sports restrictions	Yes
Ackerman	Michael	Yes, unless there is a contraindication such as asthma	Yes, LCSD	No sports restrictions	Yes

TABLE I Case 1: Additional comments (limit 100 words)	
Zareba	
Wilde	My answer to the 3rd question is provided he is on adequate beta blocker dose. Information that lacks is the QTc during exercise.
Towbin	
Sy	I would discuss LCSD if he is truly intolerant of beta-blocker, but it is not mandatory and it is contingent on local surgical expertise. The discussion about competitive sports should involve shared-decision making with patient and parents. The mode of repeat genetic testing depends on details of original testing.
Shoemaker	My decision on the sports recommendation would be personalized based on the specific activity and whether he was tolerating beta-blockers. I did not select "all sports" because I think there are lower intensity sports he could participate in safely, if he desired. A shared decision making approach is important here.
Shimizu	
Schwartz	Among genotype-negative athletes it is extremely important to rule out the possibility that adequate detraining normalizes repolarization (see Dagradi et al, Circulation in press), as this points to an exercise-induced LQTS-like pattern.
Schulze-Bahr	Information of QTc during exercise is useful for assessment.
Roden	test proband not patient. ECG morphology can be helpful (or occasionally not helpful) in directing choices e.g. if suspicious for LQT2, then less concerned about competitive sports. also circumstances under which proband presented (sleep, exercise, etc.)
Roberts	
Priori	For sport participation I would do something not considered in the list: I would suggest an implantable loop and let him participate in competitive sport on BB and monitor him with the loop recorder
Perez	The sports participation question is much more nuanced. Our approach is not a black/white restrict vs. no restrict, but rather a discussion of risks of SCD with competitive sports participation and then shared decision making. Also, a lengthy discussion of safe practices if sports participation is sought.
Lubitz	There is substantial uncertainty as to the proper management of patients with genotype negative long QT syndrome. Additionally, exercise recommendations are controversial. Common sense measures in any patient at risk for sudden cardiac arrest include avoiding exercising alone, exercising in an environment that is accessible to bystanders, and avoiding swimming and diving as a primary means of obtaining exercise. In addition, risk of exercise is likely mitigated by compliance with beta blockade, but data are scarce as noted for genotype negative long QT syndrome.
Krahn	sports participation is predicated on beta blocker adherence

<b>Kaufman</b>	
<b>Etheridge</b>	All answers assume we are convinced this boy has LQTS after the testing
<b>Eckhardt</b>	Liberal athletic participation is assuming the individual can take beta blocker and treatment escalation is also assuming that the individual actually has LQTS, which is not clear with the data provided.
<b>Culter</b>	It would be helpful to know more details about the family history of LQTS. Specifically, is there hx of SCA and if so, circumstances surrounding the SCA event. This information would be important in having a shared decision discussion with the patient and his family regarding sport participation and more advanced therapies such as LCSD and ICD. Finally, though not a first-line option, some consideration of the risk/benefit of Metoprolol (asthma present) vs more invasive advanced therapies (ICD, LCSD) would be beneficial.
<b>Chung</b>	
<b>Cerrone</b>	Q3: I would allow sport participation only if he is tolerating adequate dose of betablocker and is compliant. Without betablocker on board I would limit to recreational activities. I would in any case discuss competitive sport participation again once in high school and college and if he is still interested reassess risk profile then. Q4: I would suggest repeating genetic test if done in a research lab based on availability of the old report for direct review; known volume of the research lab and technique used. A new test has also the advantage of allowing for del/dup analysis which was rarely available then.
<b>Behr</b>	Sports restrictions only if he cannot tolerate any medical therapy. If he cannot tolerate any betablocker then I would try mexiletine before considering denervation.
<b>Aziz</b>	
<b>Ackerman</b>	For the vast majority of cases with this case stem, the correct answer is OVERDIAGNOSIS of LQTS in the proband in the first place. So, the most important point from this case scenario is to carefully adjudicate the case to see if this patient really even has LQTS in the first place.

Voting questions Case 2:	
Q1. Would you start him on a betablocker at the time of the first visit?	1) Yes, up-titrating nadolol to maximally tolerated dose 2) No, given his low baseline heart rate and the unclear/remote history of symptoms. 3) No, a patient with depression should not be given a beta blocker. 4) No, a patient with a component of drug-induced LQTS should not be bradycardic.
Q2. If the patient does not tolerate either Nadolol or Propranolol, what would you do next?	1) Try a selective betablocker 2) Recommend an ICD 3) Recommend a LCSD 4) I would defer further treatment given his age, gender and lack of recent worrisome symptoms 5) I would not have started beta-blocker to begin with
Q3. Nadolol is introduced and tolerated. His QTc and resting heart rate remain 570msec and 50bpm, respectively. Do you believe more invasive intervention is indicated?	1) No. 2) Yes. I would recommend LCSD. 3) Yes. I would recommend a single chamber ICD. 4) Yes. I would recommend a dual chamber ICD (for intentional atrial pacing and avoidance of long short intervals secondary to AV Wenckebach).

TABLE II		First Name	CASE 2, QUESTION 1	QUESTION 2	QUESTION 3
Last Name					
Zareba	Wojciech		Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	No
Wilde	Arthur		Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	No
Towbin	Jeffery		Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	No
Sy	Raymond		Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	No
Shoemaker	Benjamin		Yes, up-titrating nadolol to maximally tolerated dose	Recommend a LCSD	No
Shimizu	Wataru		Yes, up-titrating nadolol to maximally tolerated dose	I would defer further treatment given his age, gender and lack of recent worrisome symptoms	No
Schwartz	Peter		Yes, up-titrating nadolol to maximally tolerated dose	Recommend a LCSD	No
Schulze-Bahr	Eric		Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	No
Roden	Dan		Yes, up-titrating nadolol to maximally tolerated dose	I would defer further treatment given his age, gender and lack of recent worrisome symptoms	No
Roberts	Jason		Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	Yes, I would recommend a dual chamber ICD (for intentional atrial pacing and avoidance of long short intervals secondary to AV Wenckebach)
Priori	Silvia		Yes, up-titrating nadolol to maximally tolerated dose	Recommend an ICD	No
Perez	Marco		Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	No
Lubitz	Steven		Yes, up-titrating nadolol to maximally tolerated dose	Recommend an ICD	No
Krahn	Andrew		Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	No
Kaufman	Elizabeth		Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	No
Etheridge	Susan		Yes, up-titrating nadolol to maximally tolerated dose	Recommend a LCSD	No
Eckhardt	Lee		Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	No

Culter	Michael	Yes, up-titrating nadolol to maximally tolerated dose	I would defer further treatment given his age, gender and lack of recent worrisome symptoms	No
Chung	Mina	Yes, up-titrating nadolol to maximally tolerated dose	Recommend an ICD	Yes, I would recommend a dual chamber ICD (for intentional atrial pacing and avoidance of long short intervals secondary to AV Wenkebach)
Cerrone	Marina	Yes, up-titrating nadolol to maximally tolerated dose	Try a selective beta blocker	No
Behr	Elijah	Yes, up-titrating nadolol to maximally tolerated dose	Recommend an ICD	Yes, I would recommend a dual chamber ICD (for intentional atrial pacing and avoidance of long short intervals secondary to AV Wenkebach)
Aziz	Peter	Yes, up-titrating nadolol to maximally tolerated dose	Recommend a LCSD	No
Ackerman	Michael	Yes, up-titrating nadolol to maximally tolerated dose	Recommend a LCSD	Yes, I would recommend LCSD

Case 2: Additional comments (limit 100 words)	
Zareba	
Wilde	
Towbin	
Sy	The only selective beta-blocker I would try is Bisoprolol. I would avoid Metoprolol and Atenolol. Invasive therapy would need to be considered if he develops any arrhythmic symptoms while on beta-blocker therapy.
Shoemaker	Despite being arguably asymptomatic, if beta-blockers are not tolerated and his QTc is 570ms, I would consider LCSD. My thoughts are that with a QTc that long at baseline he has little room for any secondary causes of QT prolongation that may eventually occur. For example, electrolyte disturbances from gastroenteritis, need for other mild or moderate QT prolonging medicines (e.g. antibiotics), etc. LCSD may be better tolerated than an ICD in a patient with severe depression because I think it may have less effect on QOL long-term.
Shimizu	
Schwartz	LQT1 males without symptoms off therapy by age 30, are at very low risk of having a first event. However, this is true for QTc not extremely long as in the present case. Given his QTc of 570 ms I would prefer to increase his antiarrhythmic protection with an intervention that does not impact on his quality and does not interfere with his medications.
Schulze-Bahr	
Roden	there's a role for shared decision making here: ?change antidepressant. How worried is he? 570 msec is pretty worrisome, but age 60 with no symptoms is reassuring. Make sure he doesn't add any other QT prolonging drugs. Also get old ECGs if possible.
Roberts	
Priori	
Perez	
Lubitz	Assuming the patient does not tolerate a low-dose nonselective beta blocker owing to fatigue or bradycardia, I would not attempt another beta blocker. Given the marked QTc prolongation, the patient is at elevated risk for an adverse event. I would recommend a dual chamber ICD to enable atrial pacing and administration of a beta blocker. If alternatively he tolerates a beta blocker then I would advise that, in the absence of arrhythmia, administration of a beta blocker alone is reasonable. An ICD would further lower risk but at an increased risk of adverse events related to the ICD. The decision not to proceed with an ICD would involve shared decision making and close surveillance.
Krahn	
Kaufman	
Etheridge	
Eckhardt	Escalation of therapy depends on symptoms.
Culter	
Chung	
Cerrone	
Behr	With extreme QTc prolongation there will be significant risk of TdP if his K drops or he has excessive pauses - Betablocker plus ICD seem a reasonable option if he can tolerate a betablocker.
Aziz	Feel that the progression of question stems lead us toward BB therapy.
Ackerman	LCSD is "near-curative"/"most effective" for patients with LQT1. The most important steps for this patient are not discussed and that is to closely interact with his mental health care team and review options including a brief transition to a non-QT prolonging antidepressant in order to determine how much of his current QTc is drug-related and how much is his LQT1 substrate.

Voting questions Case 3:	
Q1: Case 3, this patient (asymptomatic LQTS2, QTc 500msec, 10th week of pregnancy, mild asthma) should be treated with a beta blocker immediately after timely appropriate investigations (ambulatory cardiac rhythm monitoring monitoring, exercise test).	<ol style="list-style-type: none"> <li>1. Yes</li> <li>2. No</li> <li>3. No, treatment should be started after the baseline ECG (with QTc 500) and these tests can follow the start of treatment.</li> </ol>
Q2: Case 3, this patient (asymptomatic LQTS2, QTc 500msec, 10th week of pregnancy, mild asthma) should be treated with scaled-up therapy (LCSD, ICD)	<ol style="list-style-type: none"> <li>1. Yes, LCSD</li> <li>2. Yes, ICD</li> <li>3. Yes, both</li> <li>4. No</li> </ol>
Q3: Case 3, this patient (asymptomatic LQTS2, QTc 500msec, 10th week of pregnancy, mild asthma), assuming there are no obstetrical indications for Caesarian section, should have special care during delivery	<ol style="list-style-type: none"> <li>1. Yes, special surveillance including telemetry (vaginal delivery)</li> <li>2. Yes, special surveillance but without telemetry (vaginal delivery)</li> <li>3. Yes, Caesarian section is preferable</li> <li>4. No</li> </ol>

TABLE III		CASE 3, QUESTION 1	QUESTION 2	QUESTION 3
Last Name	First Name			
Zareba	Wojciech	Yes	No	Yes, Caesarian section is preferable
Wilde	Arthur	Yes	No	Yes, special surveillance but without telemetry (vaginal delivery)
Towbin	Jeffery	Yes	No	Yes, special surveillance including telemetry (vaginal delivery)
Sy	Raymond	No, treatment should be started after the baseline ECG (with QTc 500) and these tests can follow the start of treatment.	No	Yes, special surveillance including telemetry (vaginal delivery)
Shoemaker	Benjamin	No, treatment should be started after the baseline ECG (with QTc 500) and these tests can follow the start of treatment.	No	Yes, special surveillance including telemetry (vaginal delivery)
Shimizu	Wataru	Yes	No	Yes, special surveillance including telemetry (vaginal delivery)
Schwartz	Peter	Yes	No	Yes, Caesarian section is preferable
Schulze-Bahr	Eric	Yes	No	Yes, special surveillance including telemetry (vaginal delivery)
Roden	Dan	No, treatment should be started after the baseline ECG (with QTc 500) and these tests can follow the start of treatment.	No	Yes, special surveillance including telemetry (vaginal delivery)
Roberts	Jason	Yes	No	Yes, special surveillance including telemetry (vaginal delivery)
Priori	Silvia	Yes	No	Yes, special surveillance including telemetry (vaginal delivery)
Perez	Marco	No, treatment should be started after the baseline ECG (with QTc 500) and these tests can follow the start of treatment.	No	Yes, special surveillance including telemetry (vaginal delivery)
Lubitz	Steven	No, treatment should be started after the baseline ECG (with QTc 500) and these tests can follow the start of treatment.	No	Yes, special surveillance including telemetry (vaginal delivery)
Krahn	Andrew	Yes	No	Yes, special surveillance including telemetry (vaginal delivery)
Kaufman	Elizabeth	Yes	No	Yes, special surveillance including telemetry (vaginal delivery)
Etheridge	Susan	Yes	No	Yes, special surveillance including telemetry (vaginal delivery)
Eckhardt	Lee	Yes	No	Yes, special surveillance including telemetry (vaginal delivery)

<b>Culter</b>	<b>Michael</b>	<b>Yes</b>	<b>No</b>	<b>Yes, special surveillance including telemetry (vaginal delivery)</b>
<b>Chung</b>	<b>Mina</b>	<b>No, treatment should be started after the baseline ECG (with QTc 500) and these tests can follow the start of treatment.</b>	<b>No</b>	<b>Yes, special surveillance including telemetry (vaginal delivery)</b>
<b>Cerrone</b>	<b>Marina</b>	<b>No, treatment should be started after the baseline ECG (with QTc 500) and these tests can follow the start of treatment.</b>	<b>No</b>	<b>Yes, special surveillance including telemetry (vaginal delivery)</b>
<b>Behr</b>	<b>Elijah</b>	<b>Yes</b>	<b>No</b>	<b>Yes, special surveillance including telemetry (vaginal delivery)</b>
<b>Aziz</b>	<b>Peter</b>	<b>Yes</b>	<b>No</b>	<b>Yes, special surveillance including telemetry (vaginal delivery)</b>
<b>Ackerman</b>	<b>Michael</b>	<b>Yes</b>	<b>No</b>	<b>Yes, special surveillance including telemetry (vaginal delivery)</b>

<b>Case 3: Additional comments (limit 100 words)</b>	
<b>Zareba</b>	
<b>Wilde</b>	
<b>Towbin</b>	
<b>Sy</b>	
<b>Shoemaker</b>	
<b>Shimizu</b>	
<b>Schwartz</b>	This woman is unlikely to have many more pregnancies; that's why I would not object to CS. I would monitor her for asthma recurrences but she would be safer on beta-blockers than without them.
<b>Schulze-Bahr</b>	
<b>Roden</b>	
<b>Roberts</b>	
<b>Priori</b>	
<b>Perez</b>	
<b>Lubitz</b>	The patient has long QT syndrome and a prolonged QTc. A beta blocker can be started immediately. Delaying initiation of a beta blocker to obtain cardiac rhythm monitoring or an exercise test will not alter management, and merely prolongs initiation of preventive therapy. The risk of arrhythmia is low, particularly if taking a beta blocker, during delivery. Nevertheless, telemetry would enable closer monitoring and potentially a more rapid response should an adverse event occur during the peripartum period and would be reasonable if available and feasible.
<b>Krahn</b>	answer to question 1 is predicated on timely access to testing (exercise)
<b>Kaufman</b>	
<b>Etheridge</b>	telemetry especially after the delivery
<b>Eckhardt</b>	
<b>Culter</b>	
<b>Chung</b>	
<b>Cerrone</b>	Even if LQT2 women experience increased arrhythmic risk mostly in the postpartum months, still there is a moderate risk of arrhythmias during the labor&delivery phase, so it is advisable in my opinion having the cardiology/cardiovascular genetic team alerted and available. Telemetry and iv line should be part of the delivery care.
<b>Behr</b>	If the tests are done on the day it is best to have a baseline.
<b>Aziz</b>	Perhaps worth clarifying what is meant by special care/surveillance
<b>Ackerman</b>	All too often, this type of patient is treated incorrectly with metoprolol (since this BB has earned status as most OB-preferred BB) and then during the postpartum period, this LQT2 patient has an "on-therapy" sentinel event. Reality - metoprolol should NEVER be used for LQTS. Also, way too many Caesarian sections being done simply because of maternal LQTS status. This needs to stop.

Voting questions Case 4:	
Q1: In this Case 4 (questionably symptomatic LQT2 male with asthma and QTc 485ms), would you insist on starting a beta blocker?	<ol style="list-style-type: none"> <li>1. Yes</li> <li>2. No</li> </ol>
Q2: If so, what kind of beta blocker would you give:	<ol style="list-style-type: none"> <li>1. Nonselective beta blocker (nadolol)</li> <li>2. Selective beta-1 blocker (bisoprolol)</li> <li>3. Selective beta-1 blocker (metoprolol)</li> <li>4. I would not give him a beta blocker</li> </ol>
Q3: If he cannot take beta blocker, would you offer him mexiletine?	<ol style="list-style-type: none"> <li>1. Yes</li> <li>2. No</li> </ol>
Q4: If he cannot take beta blocker or mexililtine, would you offer him LCSD?	<ol style="list-style-type: none"> <li>1. Yes</li> <li>2. No</li> </ol>
Q5: Would you consider device extraction?	<ol style="list-style-type: none"> <li>1. No, extraction is too risky for the benefit</li> <li>2. Yes, if he still wishes it after counselling</li> <li>3. Maybe, only if he tolerates beta blocker</li> <li>4. Replacement with an S-ICD</li> </ol>

TABLE IV						
Last Name	First Name	CASE 4 QUESTION 1	QUESTION 2	QUESTION 3	QUESTION 4	QUESTION 5
Zareba	Wojciech	Yes	Selective beta-1 blocker (bisoprolol)	Yes	No	Yes, if he still wishes it after counseling
Wilde	Arthur	Yes	Nonselective beta blocker (i.e. nadolol)	Yes	Yes	Yes, if he still wishes it after counseling
Towbin	Jeffery	Yes	Nonselective beta blocker (i.e. nadolol)	Yes	No	Maybe, only if he tolerates beta blocker
Sy	Raymond	Yes	Nonselective beta blocker (i.e. nadolol)	No	No	Maybe, only if he tolerates beta blocker
Shoemaker	Benjamin	Yes	Selective beta-1 blocker (metoprolol)	No	Yes	Yes, if he still wishes it after counseling
Shimizu	Wataru	No	I would not give him a beta-blocker	No	No	No, extraction is too risky for the benefit
Schwartz	Peter	Yes	Nonselective beta blocker (i.e. nadolol)	No	Yes	No, extraction is too risky for the benefit
Schulze-Bahr	Eric	Yes	Selective beta-1 blocker (metoprolol)	Yes	No	Maybe, only if he tolerates beta blocker
Roden	Dan	No	Nonselective beta blocker (i.e. nadolol)	Yes	No	Maybe, only if he tolerates beta blocker
Roberts	Jason	Yes	Nonselective beta blocker (i.e. nadolol)	Yes	Yes	Maybe, only if he tolerates beta blocker
Priori	Silvia	Yes	Nonselective beta blocker (i.e. nadolol)	No	No	No, extraction is too risky for the benefit
Perez	Marco	Yes	Nonselective beta blocker (i.e. nadolol)	No	Yes	Yes, if he still wishes it after counseling
Lubitz	Steven	Yes	Nonselective beta blocker (i.e. nadolol)	Yes	No	Maybe, only if he tolerates beta blocker
Krahn	Andrew	Yes	Nonselective beta blocker (i.e. nadolol)	No	No	Yes, if he still wishes it after counseling
Kaufman	Elizabeth	Yes	Nonselective beta blocker (i.e. nadolol)	Yes	No	Yes, if he still wishes it after counseling



Etheridge	Susan	Yes	Nonselective beta blocker (i.e. nadolol)	Yes	Yes	Yes, if he still wishes it after counseling
Eckhardt	Lee	Yes	Nonselective beta blocker (i.e. nadolol)	No	No	Yes, if he still wishes it after counseling
Culter	Michael	Yes	Nonselective beta blocker (i.e. nadolol)	No	No	Yes, if he still wishes it after counseling
Chung	Mina	Yes	Nonselective beta blocker (i.e. nadolol)	No	No	Yes, if he still wishes it after counseling
Cerrone	Marina	Yes	Nonselective beta blocker (i.e. nadolol)	Yes	No	Yes, if he still wishes it after counseling
Behr	Elijah	Yes	Nonselective beta blocker (i.e. nadolol)	Yes	Yes	Yes, if he still wishes it after counseling
Aziz	Peter	Yes	Nonselective beta blocker (i.e. nadolol)	Yes	Yes	Yes, if he still wishes it after counseling
Ackerman	Michael	Yes	Nonselective beta blocker (i.e. nadolol)	Yes	Yes	Maybe, only if he tolerates beta blocker

Case 4: Additional comments (limit 100 words)	
Zareba	
Wilde	
Towbin	
Sy	
Shoemaker	I would rely heavily on shared decision making with this case. While selective beta-blockers are not as effective as nadolol or propranolol, they still have some efficacy. So, if he can tolerate metoprolol or bisoprolol and has never had a breakthrough episode, I would consider removing the ICD. That would be my preferred option (if he strongly wants device removed). If he could not tolerate a beta-blocker, then LCSD and ICD removal would be my second preferred option. I would try to dissuade him from ICD removal without another SCD risk reduction measure, especially if he exercises or has other risk factors. He is likely approaching ERI (DOI 9 years ago), so the decision will ideally be made soon.
Shimizu	
Schwartz	The initial decision to implant the ICD was not supported by published data or guidelines.
Schulze-Bahr	
Roden	shared decision making again
Roberts	
Priori	The last question: Since he has an ICD that provides best protection I would recommend BB according to guidelines, but I would focus my conversation with him to remain on ICD (without BB) rather than insisting to add BB and removing ICD
Perez	I would counsel strongly against extraction of lead given high risk. That said, if patient is determined to have device extracted and is counseled appropriately, then I would consider it. Another option is to cap the lead without extracting and remove the generator.
Lubitz	
Krahn	response to question 1 assumes a selective BB can be started in the non selective one is not tolerated
Kaufman	
Etheridge	
Eckhardt	Nonselective beta blocker trial is reasonable but if asthma worsens then would choose selective beta blocker. Shared decision making around ICD explant is essential and counseling on the importance for compliance with medical therapy and avoidance of QT prolonging medications is part of that decision. Additionally, explant within 10 years of initial implant is preferred as lead systems older than 10 years (particularly dual coil systems) are a higher risk to remove. Explant recommendation is center dependent, as our experience is that our high volume for extraction significantly reduces risk of explant.
Culter	Prior to a shared decision on extraction, extensive discussion of the risks of extraction is needed. In particular, given the young age of ICD implant it is possible that as the patient grew the lead may have become imbedded into the vessel wall of the SVC, increasing the risk of SVC tear. Pre-procedure advanced imaging could be considered to evaluate how the lead interacts with the vessel wall.

<b>Chung</b>	
<b>Cerrone</b>	<p>Q2-Q3: I would try different beta blockers before offering mexiletine. I would also monitor if mexiletine therapy (once decided it is needed) causes a shortening of the QTc.</p> <p>Q5: I would reinforce that device extraction has to be performed only in centers with high volume of extraction procedures and known expertise. Careful avoidance of all QT-prolonging medications and prompt correction of other triggers (hypokalemia etc) should also be strictly reinforced. LCSD could be discussed if he cannot tolerate any medical therapy and opts for device extraction.</p>
<b>Behr</b>	
<b>Aziz</b>	
<b>Ackerman</b>	<p>Bottom Line: even though asymptomatic so far, LQT2 + his QTc compels an intentional LQT2-directed treatment program. I would not explant his ICD system until confident that such a program (BB or BB + mexiletine or mexiletine only or LCSD or something else) has been put in place successfully. Then proceed with device removal.</p>