

Idebenone Survey
March 2005

| Age | Weight | Gender | Age at onset or Dx | Time on Idebenone | Dosage (mg/kg/d) | Other meds, supplements | Side effects noted | HCM involved? | Reduction or stabilization of HCM? | Non-cardiac symptom changes? | Other observations, comments | Where purchased | Cost | Dosage form and size | Can child swallow pills? | Alternate administration | Interested in group purchase? | Doctor comments |
|-----|--------|--------|--------------------|-------------------|------------------------------|--|--------------------|---------------|--|--|---|-------------------------------------|---------------------------|---------------------------------|--------------------------|--------------------------|-------------------------------|--|
| 33 | 110 | F | Dx 22 | 5-6 yrs | 6.3 (7x45 mg/d) | Vit E | none | no | | less tired | Our daughter's cardiologist thinks that taking idebenone will prevent her from having heart problem. (a french study has shown the real good effect of idebenone on most children with cardiomyopathy). On top of that, it gives her energy. (that is my opinion) | French govt provides | 0 | 45 mg tab | yes | | no | nothing special except to do exercises to remain as fit as possible until a new medicine is found to cure FA |
| 12 | 75 | M | 5 (Dx 7) | 5 mo | 10 | Vit E 400 IU | none | yes | don't know yet | no | More energy, tires less easily, feels better happier. Sometimes a little hyper. | Smart Drugs | \$80/mo. | 150 mg cap 45 mg cap | yes | | yes | I had mentioned Idebenone 2 years ago to XXX's neurologist, she said she would look at it but recommended CoQ10 with Vitamin E. I did some research and found NIH (National Institute of Health) signed XXX's name up and we were contacted about the study. Once he started the study at NIH and I seen how much better he felt and acted, I knew I had to get him on Idebenone. My thing with XXX's doctor is she tells one thing to one family and then something to another. Dr. Wong told me she would help me monitor XXX's Idebenone and yet that same day or week told another parent she couldn't do the. She leaves me confused and frustrated. I do know I can call Dr. Di at NIH and he will have a test sent right away if I feel something is wrong. I think I am going to see how his cardiologist is feeling about the med and see what I can do from there. Either way XXX will continue to take it and unless I get some huge bad blood test report back he is on it for good. |
| 28 | 155 | M | 9 (Dx 15) | 9 mo | 5 | CoQ10 600mg/d previous to Idebenone, NAC 2400mg, Vit E 800 IU, Vit C 500 mg, Multivitamin w/o Iron, Calcium, Fiber | none | very mild | stable since Dx | no | repeats \$25/925 | Kirkman | \$68/mo. | 150 mg cap | yes | | yes | Nothing, but our primary care doctor has been supportive and willing to let us try all of these. Cardiologist say he is not sure it will help (he was not up on idebenone research I asked if it would hurt, and he said he did not believe that it would hurt. |
| 13 | 115 | F | 13 (Dx 5) | 9 mo | 10 | CoQ10 600mg/d previous to Idebenone, NAC 2400mg, Vit E 800 IU, Selenium 50 mcg, Evening primrose oil 1300mg, famotidine (Pepcid AC) 10mg as acid reducer | none | very mild | stable since Dx | no | repeats 1150/820 CoQ10 helped energy level | Kirkman | \$102/mo. | 150 mg cap | yes | | | |
| 27 | 105 | F | 9 | 3 yrs | 10 | | none | no | | no | no | Kirkman | \$0.50/cap | 150 mg cap | yes | | already do | nothing |
| 8 | 59 | M | 5 (Dx 8) | 3 mo | 10 | multivitamin, Vit E 800 IU | none | yes | increased from 12 to 15 mm from Sept to March | | balance seems better | Smart Nutrition | 12 boxes of 60 caps \$240 | 45 mg cap | no | applesauce | yes | Our doctors refuse to talk about Idebenone and act like they've never heard of it. |
| 14 | 105 | F | 7 (Dx 10) | 3.5 yrs | 5 | Vit E 1200 IU, selenium, alpha lipoic acid, L-carnitine, folic acid, psych meds | none | no | | falls ceased on start, still walking, recuperated well after spinal fusion | CoQ10 not nearly as effective. Symptoms worsen if she goes off Idebenone. | Kirkman | 12 bottles \$320 | 45 mg tabs | yes | | yes | Our neurologist (an MDA doc) asks us every time we go where we get it, always seems surprised when we repeat that we don't have to get it from Europe, but he seems to be aware that there is good news about the promise of Idebenone treatment. He is very impressed with how well Bridget is doing, so far. So are we! |
| 21 | 140 | M | 16 | 8 mo | 5 (6mo) 10 (2 mo) | Atenolol | none | yes | no change | no | no | Smart Nutrition | 12 boxes of 30 caps \$336 | 150 mg cap | yes | | yes | Nuero recommended 300 mg Co-Q10, Vit E. He has no knowledge of Idebenone |
| 14 | 75 | F | 13 | 2 wks | 8.8 | Vit E, multivitamin | none | mild | don't know yet | | | local pharmacy through Smart Source | \$24/pkg | 150 mg cap | yes | | not now | MDA doctors don't encourage us taking it, family doctor who has had a child with cancer, in remission now, say's he understands and is willing to help us try whatever we would like, as long as it doesn't cause a problem! |
| 14 | 91 | M | 8 (Dx 13) | 1 yr | 5 (2 mo) 15 (9 mo) 18 (1 mo) | Vit E 800 IU (400 alpha tocopherol succinate, 400 "complete E" with all 8 forms of E) | none | yes | mild initially, slight increase at 6 mos., started to reverse at 12 mos. | | scoliosis | Kirkman | \$0.57/cap | 150 mg cap | yes | | yes | We only see Drs who are supportive of our desire to do everything possible to slow the disease progression of our son. The first neurologist wasn't so we don't see him anymore. |
| 8 | 50 | M | 6 | 2 yrs | 5 1st yr 7.5 last yr | CoQ10 and Vitamin C | none | no | heart on high end of normal range | no | energy increases after starting Idebenone and then also after starting CoQ10 | Smart Nutrition | \$29.95 for 60 | 45 mg tab | yes | | sure | The MDA clinic still sticks to the traditional Co-Q, however my Neurologist at Columbia University's Movement disorders clinic, supports Idebenone (more soluble). He also is watching carefully the development of the Mito-Q. We just had our Neuro appointment and I had the courage to ask if he saw any difference, and the answer was no for the ataxia, and is more concerned with watching the Heart and spine. |
| 11 | 90 | F | 4 (Dx 8) | 3 yrs | 5 (1.5 yrs) 10(1.5 yrs) | CoQ10 300 mg, Vit E 800 IU, selenium 200 mcg, L-carnitine 500 mg, alpha lipoic acid 120 mg, fish oil 1000 mg, atenolol (beta blocker) 37.5 mg | none | yes | severe, 16mm up to 18mm at age 8-9, shrinking since idebenone. Now at 9mm. | no | repeats 900/1200 More energy since heart is improving, but hard to know whether beta blocker is contributing to that. | Kirkman, through sherywys.com | \$24.99 for 60 | 45 mg cap, moving to 150 mg cap | yes | | yes (already do) | XXX's cardiologist was involved in an early trial of idebenone in Texas and saw positive results. He is thrilled to see her progress. |
| 14 | 120 | M | 13 | 8 mo | 13.6 (750 mg/d) | CoQ10 300mg, Vit E 800 IU, 1/2 multivitamin | none | slight | don't know yet | continues to get clumsier | no | Smart Nutrition | 12 boxes of 30 caps \$336 | 150 mg cap | yes | | yes | Doctor has not recommended anything. |