An Innovative Collaborative Model of Care for Undiagnosed Complex Medical Conditions

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A recent National Academy of Sciences report titled Improving Diagnosis in Health Care emphasizes the importance of right diagnosis in health care delivery. Missed or delayed diagnoses, or incorrect diagnoses, constitute a patient safety problem. Diagnostic errors may contribute to ~10% of patient deaths and account for a large portion of malpractice claims.

One reason for diagnostic errors is the lack of collaboration between clinicians. Collaboration is particularly problematic when clinicians involved in the child’s care practice in different settings. The National Academy of Sciences report advocates teamwork in the diagnostic process. Collaborative, interdisciplinary conferences are often used in health care settings to improve patient care. Tumor boards, conferences where clinicians come together to discuss plans of care for their patients with cancer, improve patient outcomes. However, collaborative models of care to make diagnoses are not generally available. We describe an innovative model of care for children with undiagnosed complex medical conditions (UCMC) developed in a tertiary care children’s hospital as a result of family partnership with a health care system.

CONTEXT (NICOLE DONOGHUE)

Declan was a wonderful baby. He was outgoing, sweet, and talkative. He had a wonderful focus and a beautiful spirit and was always happy and smiling as long as he was playing. Unlike most children, Declan had another list of details that described him. Declan’s medical problem list included fevers, eye scarring, arthritis of the knees, liquid aspiration, lower lobe pneumonia, erratic and nonconclusive blood test results, and unexplained illnesses.

Declan had 10 specialists working to solve the mystery surrounding his unexplained illnesses. Each physician, including his primary care physician (PCP) and multiple specialist physicians, tried to solve our medical questions through their singular lens. But there was no 1 person trying to piece it all together. I prepared a medical overview chart documenting the outcomes of every visit and hoped to make some kind of connection between his ailments and symptoms. With >100 e-mails that I sent and coordinated between specialists, I was in the middle of the specialists without satisfactory results. What I really needed was a medical quarterback helping me navigate all the results, the assumptions, and our options. But Declan did not have a medical quarterback; he had a tenacious mom unwilling to let her son’s ailments go unanswered. But we ran out of time. The fevers got too high and too low and we got desperate for answers. At the age of 2, Declan was diagnosed with juvenile idiopathic arthritis and treated with prednisone. After 2 days of treatment, Declan died from overwhelming sepsis.

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Dr Nageswaran conceptualized and designed the study, drafted the initial manuscript (except the Context section), analyzed the data, and reviewed and revised the Context section; Ms Donoghue drafted the Context section and reviewed and revised the other sections; Ms Mitchell analyzed the data and reviewed and revised the manuscript; Dr Givner reviewed and revised the manuscript; and all authors approved the final manuscript as submitted.

DOI: 10.1542/peds.2016-3373

Accepted for publication Feb 16, 2017

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PEDIATRICS (ISSN Numbers: Print, 0031-4005; Online, 1098-4275).

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FINANCIAL DISCLOSURE: Dr Nageswaran is the director of the Pediatric Enhanced Care Program of Brenner Children’s Hospital, which has received financial gifts from the Declan Donoghue Foundation, and Ms Donoghue, Ms Mitchell, and Dr Givner have indicated they have no financial relationships relevant to this article to disclose.

FUNDING: No external funding.


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After his death, Declan was diagnosed with the rare genetic disease, X-linked chronic granulomatous disease. Declan’s father and I reviewed the many communications we had with Declan’s doctors and identified the missed opportunities to correctly diagnose Declan’s condition primarily from lack of communication between the many care providers and a clinician or team that served as a quarterback, overseeing his care. We articulated to the leaders of Wake Forest Baptist Health (WFBH) our concerns about the gaps in the process of diagnosing Declan. We then worked with WFBH leaders to create a system to address these gaps in making diagnoses for children. This partnership led to the development of the Declan Donoghue Collaborative Care Program (DDCCP) in April 2011.

PROGRAM (SAVITHRI NAGESWARAN, ANGEL MITCHELL, AND LAURENCE GIVNER)

Description

The goal of the DDCCP is to reduce morbidity and mortality in children with UCMC or complex medical conditions with unresolved clinical problems. Before the program started, a literature review was done to identify already developed collaborative models of care. An e-mail inquiry was sent to the complex care distribution list in the country. There were descriptions of interdisciplinary meetings; however, no other programs such as the one that was envisioned existed. A preliminary proposal for the program was developed, sent to all the physicians caring for children at WFBH, and then revised based on their feedback. The program model was continually revised based on the experiences in project implementation.

DDCCP is a program within the pediatric palliative care and complex care program of Brenner Children’s Hospital (BCH), a tertiary care children’s hospital of the WFBH system in western North Carolina. DDCCP personnel include a 0.15 full-time equivalent pediatrician and a 0.5 full-time equivalent master’s-level registered nurse who are the program director and the coordinator, respectively. WFBH provides funding for personnel for the program. Referrals to DDCCP can be initiated by the child’s PCP or specialist physician. To be eligible for the program, the child must already be receiving care at BCH, must have an undiagnosed clinical condition or unresolved clinical problem after a workup was done, and should need or receive care from ≥3 specialists. Exclusion criteria are referral only for care coordination, social or psychological reasons, and urgent clinical situations.

Once the child is deemed eligible, the coordinator contacts the child’s parent or caregiver, describes the program, and obtains his or her permission to enroll the child in the program. The next step in the DDCCP process is when the coordinator, using a format that Declan’s mother created to organize his clinical information, summarizes all medical information from records at BCH and all outside clinicians from whom the child received care. This comprehensive clinical summary includes information about all clinic (primary care and specialist) and emergency department visits, hospitalizations, and results of laboratory tests and procedures (see Supplemental Materials). Significant findings and abnormal values are color coded.

Next, the coordinator obtains a detailed history from the parents, including their concerns about their child’s condition, goals for the child, current medications, and family history; these data are added to the summary. The program director reviews the summary and, if necessary, obtains more clinical information from the parents or conducts a clinic visit. The final version of the clinical summary is placed in the child’s electronic medical record (EMR).

An invitation to participate in a meeting in person or by phone is sent to all clinicians involved in the child’s care, including nonphysician clinicians, the child’s PCP, and clinicians whose input the DDCCP team or the referring physician thinks will be valuable. For those unable to join the meeting, the program director obtains their input beforehand.

Meetings are held after clinic hours on weekdays and last for ~75 minutes. Before the meeting, the program director sends the comprehensive clinical summary and questions to be discussed at the meeting to all participants. These discussion questions are generated based on parents’ or referring physicians’ concerns and the review of the child’s summary, and they are used as a guide to facilitate the meeting. Because the clinical summary is shared with clinicians ahead of time, participants have the opportunity to review the summary before the meeting.

All ideas generated in the meeting are captured in a detailed written report, including the rationale for excluding or including diagnoses. A plan is developed with clearly identified action items including additional tests to be done, specialist opinions to be sought, communication plans, and the person responsible for each action item. This meeting report is communicated with parents, shared with all clinicians involved in the child’s care, and placed in the child’s medical record.

Next, the DDCCP team works on implementing the plan and oversees the child’s care until a diagnosis is
Outcomes

A database of referral data, child characteristics, conference information, and outcomes is maintained by the DDCCP team. Descriptive statistics (medians and proportions) were used to summarize data. Institutional review board approval was obtained to allow presentation of these data.

From April 2011 through June 2016, the DDCCP received 66 referrals. Before a summary was developed, a diagnosis was reached in 4 cases, 5 withdrew from the program, 5 were deferred because it was subsequently determined that a conference was not necessary, and 4 are in the early phase and have not yet had a summary developed. Of the remaining 48 children for whom a summary was developed, in 6 cases a diagnosis was reached before the conference was conducted, 1 withdrew from the program, and 1 was deferred because a conference was deemed unnecessary by a specialist physician. Conferences were held for 40 children (24 boys and 16 girls). The median age was 5 years (range 1 month–17 years). For 29 children, the conference was initiated to find an underlying diagnosis, and for 7 it was to develop a plan of care. The median number of specialists involved in the care of these 36 children was 7 (range 3–15). In 4 cases, children had died suddenly and unexpectedly, and the conference was held to search for a diagnosis, including possible genetic disorders, and thus clarify risk for other family members and future offspring.

Of the 29 children referred for diagnosis (excluding the 4 children who were referred after their death), for 21 (72%), a diagnosis was reached, and for 8 (28%) a diagnosis was not reached (Table 1). All children without diagnoses remain in the program and are doing well clinically. Two children referred for diagnosis subsequently died. Both had rare incurable genetic disorders (dyskeratosis congenita and leukodystrophy) diagnosed through the collaborative process. A diagnosis was reached in 1 of the 4 children who had died suddenly and unexpectedly (Table 1), and a plan was reached for surviving family members of all 4 children. Plans of care were developed for all 7 children with unresolved clinical problems referred for developing a plan. One child referred for developing a plan for an unresolved clinical issue died subsequently, after bone marrow transplantation. The success of the DDCCP is illustrated by case examples in Table 2.

Lessons Learned

Clinical summaries provide clinicians with complete information about the child and enable them to discern problems easily. Before reviewing the clinical summary, many clinicians were not fully aware of the other specialist visits, testing done, or opinions. However, developing a clinical summary takes a lot of time and thus may delay the collaborative care process. EMR capabilities would be valuable, but this feature

<table>
<thead>
<tr>
<th>TABLE 1 Diagnoses and Reasons for Not Reaching Diagnosis ($n = 33$)</th>
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<tbody>
<tr>
<td>Diagnosis</td>
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<tr>
<td>Diagnosis reached</td>
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<tr>
<td>Alagille syndrome</td>
</tr>
<tr>
<td>Dyskeratosis congenita</td>
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<tr>
<td>Leigh syndrome</td>
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<tr>
<td>Primary bile acid malabsorption</td>
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<tr>
<td>Leukodystrophy</td>
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<td>Tracheal defect</td>
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<tr>
<td>Eosinophilic esophagitis</td>
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<td>Nutritional deficiency due to dysphagia</td>
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<tr>
<td>Consequences of prematurity</td>
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<tr>
<td>Drug-induced bone marrow suppression</td>
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<tr>
<td>Autonomic dysfunction disorder</td>
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<tr>
<td>Joint hypermobility syndrome</td>
</tr>
<tr>
<td>Factitious disorder</td>
</tr>
<tr>
<td>Medical child abuse</td>
</tr>
<tr>
<td>Hypertrophic cardiomyopathy$a$</td>
</tr>
<tr>
<td>No diagnosis reached has a summary developed</td>
</tr>
<tr>
<td>Additional testing not done because of lack of funds for testing</td>
</tr>
<tr>
<td>Awaiting genetic testing$b$</td>
</tr>
<tr>
<td>Awaiting test results</td>
</tr>
<tr>
<td>No diagnosis after complete workup$c$</td>
</tr>
</tbody>
</table>

a Referred after death.
b Includes 1 child referred after death.
c Includes 2 children referred after death.
**TABLE 2 Illustrative Cases of Outcomes of DDCCP**

| Example 1 | A 10-y-old girl presented with feeding intolerance; vomiting; bloody diarrhea; electrolyte imbalances; low levels of vitamins A, D, E, and K; anemia; stunted growth and height; nephrolithiasis; muscle weakness, and fracture due to osteopenia. She was receiving care from 10 specialists, hospitalized 7 times in 10 mo, in the emergency department numerous times, and wheelchair bound. A collaborative conference was held, and a plan was developed. A diagnosis of primary bile acid malabsorption was made, and treatment with a bile acid binder was started. Since then, almost all her laboratory abnormalities have resolved, and she has not been hospitalized for nearly 2.5 y. She is walking, playing, and attending school and has been in her best health in many years. |
| Example 2 | A 2.5-y-old boy was repeatedly hospitalized for feeding intolerance, abdominal pain, and need for total parenteral nutrition. He had multiple neurologic symptoms. He was dependent on methadone and hydromorphone for pain control. He was being evaluated for an intestinal transplant. In the conference to discuss his case, there were 23 participants representing 15 different specialties. Other diagnoses were excluded, and a diagnosis of medical child abuse was made. |
| Example 3 | An 11-y-old girl developed severe ascites after an episode of toxic epidermal necrolysis. She needed repeated (almost monthly) paracentesis for ascites and was hospitalized often. She had growth delay, angiookeratomas of her skin, coarse features, dystrophic nails, anemia, and mild hepatic fibrosis. A collaborative conference was held, and she was hospitalized as part of the plan to conduct additional studies. A second conference was held after this hospitalization, and a plan was developed and implemented. Based on this plan, her ascites is managed medically with diuretics. Although a diagnosis could not be made, she has not been hospitalized for 3 y and has not needed additional paracentesis. |

is currently not possible in the institution’s EMR system. However, EMR capabilities will not affect other components of the DDCCP model (eg, conference).

Clinician engagement is important. Although clinicians’ participation in the conference is a nonreimbursable clinical effort, DDCCP has maintained high clinician participation. Of those invited for a conference, 64% participated in person or by phone, and 88% provided input in some way. Clinicians participating in the conference noted that it was helpful to hear the thought processes of other clinicians, which may not be captured in the clinic visit notes. Scheduling the meetings after clinic hours and providing opportunities to join by phone increased clinician participation. Getting the opinions of clinicians who could not participate facilitated clinician engagement. Early in DDCCP development, only specialist physicians were invited to the conference. However, recognizing the importance of other clinicians in the process, the program evolved to include PCPs, clinicians from outside institutions, and nonphysician clinicians.

The importance of a team that oversees the care of the child became even more obvious as the program progressed. Because the care of UCMC is highly fragmented, a clinical team that takes responsibility for coordinating the care, ensuring implementation of all tasks identified in the conference, and maintaining communication with all relevant clinicians is very important to the collaborative model.

Arriving at a diagnosis sometimes takes many months. Lack of insurance reimbursement for some tests (eg, whole exome sequencing) is a major limitation to arriving at a diagnosis. Although some children were referred to research programs, not all children are eligible for these programs; even if they are, there is often a waiting period. Reimbursement for all appropriate testing will improve and prevent delay in diagnosis. In a small group of children, diagnosis could not be reached even after complete testing; implementing a coherent plan of care improved the health outcomes of these children.

**CONCLUSIONS**

A collaborative model of care to improve the diagnostic process can be implemented in tertiary care children’s hospitals. Although the activities of DDCCP are aligned with the medical home model, PCPs may not have the resources in their practice or elsewhere to accomplish all the activities involved in the DDCCP model. Many children’s hospitals have programs for children with medical complexity in an effort to better manage the care that they receive from multiple clinicians. However, the DDCCP is different from these programs in that it incorporates the principles of interdisciplinary conferences and care management by using a highly structured process with a singular goal of making diagnoses in UCMC.

The DDCCP helped make diagnoses, exclude diagnoses, and implement a plan of care for UCMC. A collaborative model of care such as the DDCCP can help prevent erroneous diagnoses and treatments and avoid unnecessary tests, procedures, and hospitalizations and lead to improved health outcomes for children. The DDCCP model of care for UCMC can be replicated in other institutions caring for children. It can also be a useful model in other populations (ie, adults) to decrease fragmentation of care when multiple health care providers are involved in the care of a patient. Although demonstrating cost savings was not the goal when DDCCP was started, we are
conducting a return on investment analysis. Demonstrating cost savings from DDCCP will increase the sustainability of the program. Implementation of the DDCCP is an example of how family partnerships can help a health system improve its care delivery for children. Our goal is for all UCMC to have access to a collaborative model of care in which a health care team takes responsibility for and is given the time necessary to do all the care coordination activities involved in arriving at a diagnosis.

ACKNOWLEDGMENTS
We thank Karen Klein, MA, ELS, of the Wake Forest Clinical and Translational Science Institute (UL1 TR001420; principal investigator McClain), for editing the manuscript.

ABBREVIATIONS
BCH: Brenner Children’s Hospital
DDCCP: Declan Donoghue Collaborative Care Program
EMR: electronic medical record
PCP: primary care physician
UCMC: undiagnosed complex medical conditions
WFBH: Wake Forest Baptist Health

POTENTIAL CONFLICT OF INTEREST: The authors have indicated they have no potential conflicts of interest to disclose.

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An Innovative Collaborative Model of Care for Undiagnosed Complex Medical Conditions
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Pediatrics 2017;139; originally published online April 24, 2017;
DOI: 10.1542/peds.2016-3373

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