

Managing Disagreements in the Management of Short Bowel and Hypoplastic Left Heart Syndrome

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ABSTRACT. *Background and Objectives.* When innovative, not yet fully proven therapies are introduced, physicians may have neither experience nor sufficient data in the medical literature to assist in their decision to discuss them with and/or recommend them to patients. Little is known about how physicians deal with this uncertainty. Moreover, when multiple physicians caring for a single patient have reached different conclusions regarding this new therapy, the potential for disagreement exists that could give rise to ethical issues as well as cause confusion to the patient. To explore these topics, we investigated the attitudes of specialists to therapies for two life-threatening diseases: hypoplastic left heart syndrome (HLHS) and short bowel syndrome.

Methods. A forced choice questionnaire was distributed to the heads of neonatology, pediatric cardiology, and pediatric gastroenterology training programs asking about their outcome impressions and treatment recommendations and about the local availability of treatments. In addition, responses from specialists from the same institution were linked in a confidential manner to evaluate the frequency of disagreement within the same institution. Responses were analyzed using χ^2 and Wilcoxon matched pair analysis as appropriate.

Results. The overall rate of response was 79%. In institutions that had both neonatology and pediatric gastroenterology training programs, there was a 59% response rate compared with a 73% response rate from institutions that had both neonatology and cardiology programs. Significant differences were noted among specialists as to who would be involved in discussions of therapeutic options with patients in both HLHS and short bowel syndrome. Differences also were noted in the willingness of specialists to discuss and recommend therapies, in the perceived survival and quality of life by various specialists after transplant and palliative surgery, and in the local availability of various options. The neonatologists and gastroenterologists at the same institution disagreed on responses in 34% of the questions with only 1 of the 25 pairs in full agreement. In contrast, the neonatologists and pediatric cardiologists at the same institution disagreed in only 14% of the questions with 7 of the 28 pairs in full agreement.

Conclusions. Substantial disagreement among specialists about new interventions was found. There seem

to be fewer differences among specialists when dealing with the more mature therapy, HLHS. Two major ethical issues arise. First, there seems to be no accepted professional standard to which individuals can appeal when determining whether to discuss or recommend new, not-yet-fully-proven technologies. Second, there is the potential for much patient confusion when counseling physicians recommend different options. Colleagues as individuals and specialists as groups should talk to each other before individual discussions with families to ensure that there is a clear understanding of differing beliefs. *Pediatrics* 1999;104(4). URL: <http://www.pediatrics.org/cgi/content/full/104/4/e48>; forecasting, prognosis, survival rate, quality of life, counseling, decision making, policy making, questionnaires, short bowel syndrome, parenteral nutrition, intestine, small transplantation, surgical stapling, hypoplastic left heart syndrome, heart transplantation, palliative care methods.

ABBREVIATIONS. SBS, short bowel syndrome; TPN, total parenteral nutrition; HLHS, hypoplastic left heart syndrome.

In the management of life-threatening diseases, standard therapies are those generally accepted by physicians and recommended to their patients. However, physicians also may consider newly introduced, innovative treatments of which the benefits have not yet been scrutinized fully scientifically. During this period, between the therapy's introduction and its acceptance as beneficial, managing physicians are left to: 1) assess the value of such new therapies through different avenues (personal experience, limited scientific reports, and the experience of colleagues or specialists); 2) decide in light of that assessment whether there is sufficient preliminary evidence of a favorable risk-benefit ratio to justify discussing this option with the patients and informing them of its availability; and 3) determine in light of the preliminary evidence whether it is appropriate to recommend these new therapies. These assessments, decisions, and determinations present many ethical challenges, especially when the data offer no firm guidance.¹ Little is known about how physicians deal with these challenges.

In addition, it is not uncommon to have several physicians of differing specialties simultaneously managing a patient with a life-threatening disease. These multiple physicians will agree most often in the choice of therapy when the therapy has been accepted as beneficial and has become part of the standard of care. However, significant potential for disagreement exists when physicians are considering

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Received for publication Dec 23, 1998; accepted Apr 8, 1999.

Presented in part at the Society for Pediatric Research Annual Meeting; May 4, 1998; Washington, DC.

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newer therapies that are not accepted yet as beneficial and when responsibility for decision making is shared among these specialties. This potential for disagreement gives rise to additional ethical issues, both in the relationships among these specialists and in their relationships with their patients. Little also is known about how clinicians deal with these issues.

In newborn patients, short bowel syndrome (SBS), attributable either to a primary congenital anomaly or to necrotizing enterocolitis, can be lethal. Although it does not cure the problem, long-term total parenteral nutrition (TPN) has become accepted widely as beneficial.² New surgical interventions (ie, surgical intestinal lengthening or small bowel transplantation) that offer the hope of a functioning organ that allows complete enteral nutrition have been introduced.²⁻¹¹ Although these surgeries may be deferred beyond the neonatal period, discussions of them as options may begin while these patients are cared for by both neonatologists and pediatric gastroenterologists. Uncertainties about the benefits of these surgical options combined with the potential for differences among these professionals in the clinical acceptance of these new therapies could give rise to the ethical issues mentioned above.

We undertook a study to investigate the impressions and recommendations of neonatologists and pediatric gastroenterologists in caring for patients with SBS. As part of this study, we also investigated similar impressions and recommendations of neonatologists and pediatric cardiologists regarding more mature, innovative surgical therapeutic options for hypoplastic left heart syndrome (HLHS),^{12,13} the use of which we have investigated in a previous study.¹⁴

METHODS

A forced choice questionnaire (Fig 1 and Fig 2) was distributed to the 102 heads of neonatology training programs (both questionnaires), the 47 heads of pediatric gastroenterology training programs (SBS questionnaire), and the 46 heads of pediatric cardiology training programs (HLHS questionnaire), identified by the Accreditation Council for Graduate Medical Education. Approval for the study was obtained from the institutional review board of Baylor College of Medicine.

Each institution was assigned an identifying number. A single master list was maintained linking the institution and an identifying number. That number, together with an indication of the specialty, was added to the face of each questionnaire before mailing. As each questionnaire was returned, the receipt of the response was noted on the master list. Nonrespondents were remailed the questionnaire once more. After receipt of the responses to the second mailing and before data entry and analysis, the master list linking the institutional names with the institutional numbers was destroyed to preserve the confidentiality that we promised the respondents. The institutional number was used to link responses from different professionals at the same institution to perform paired sample analysis.

Some respondents did not complete all questions. We report the number and percent of responses for each question. Responses from the same institution were compared for agreement and disagreement only if both respondents from that institution answered the question.

At the beginning of the SBS survey, we established our definition of SBS as the length of bowel deemed inadequate to allow adequate nutrition by a completely enteral route. We then queried the respondents for their own determination of this length both with and without an intact ileocecal valve and asked that they use that definition to answer additional questions.

For consistency and clarity, we established the following vocabulary for the purpose of this paper: 1) intraprofessional variance, variability of responses within a discipline (eg, variance among all neonatologists regarding outcome of SBS); 2) interprofessional difference, differences between the overall responses of the members of two professions (eg, the overall opinions of the pediatric gastroenterologists vs the overall opinions of the neonatologists about gut transplant); 3) cross-disease comparison of interprofessional difference, the degree of interprofessional difference for neonatologists/gastroenterologists as opposed to the degree of interprofessional difference for neonatologists/cardiologists (eg, do neonatologists and pediatric gastroenterologists disagree more about the outcome of gut transplant than neonatologists and pediatric cardiologists disagree about the outcome of heart transplant); 4) intrainstitutional dissonance, differing opinions among specialists in the same institution (eg, the frequency of differences in the impressions of the gastroenterologist and the neonatologist at a specific hospital about gut transplant); 5) cross-disease comparison of intrainstitutional dissonance, the degree of intrainstitutional dissonance for neonatologists/gastroenterologists as opposed to intrainstitutional dissonance for neonatologists/pediatric-cardiologists (eg, was there more intrainstitutional dissonance between neonatologists and pediatric gastroenterologists regarding the outcome of gut transplant than between neonatologists and pediatric cardiologists about the outcome of heart transplant).

This vocabulary enables us to discuss a range of possible disagreements. For example, it may be possible for the two groups of professionals to have the same average response among all respondents. In that case, there would be no interprofessional differences, but there still could be significant dissonance within institutions. Suppose that 50% of both cardiologists and neonatologists recommend treatment option A. Then there would be no interprofessional difference. However, it is still possible for there to be no agreement on treatment recommendations within any institution if the cardiologists recommending treatment option A are at one half of the institutions while the neonatologists recommending treatment option A are at the other half of the institutions. There then would be tremendous intrainstitutional dissonance.

χ^2 with Yates correction and Wilcoxon matched pair analysis were used as appropriate.

RESULTS

We surveyed the directors of the 102 neonatology, 47 pediatric gastroenterology, and 46 pediatric cardiology programs in May 1997 with a response rate of 72 of 102 (70%) for the neonatologists, 40 of 47 (85%) for the pediatric gastroenterologists, and 42 of 46 (91%) for the pediatric cardiologists. This provided us with an overall response rate of 154 of 195 (79%). We received 27 of 46 (59%) responses from the neonatology-gastroenterology pairs (there was a single gastroenterology program not associated with a neonatology program) and 33 of 46 (73%) responses from the neonatology-cardiology pairs from the same institution.

Interprofessional Differences

Although they concede that they are not the primary discussants, the majority of the neonatologists considered themselves involved in the discussions with parents about treatment options for SBS. This perception was denied by the pediatric gastroenterologists (see Table 1).

Responding neonatologists had a significantly different understanding of the length of bowel sufficient for successful enteral nutrition than did pediatric gastroenterologists. They were significantly more optimistic believing that less gut was required

- 1) **By short gut we are referring to infants with an inadequate amount of small/large bowel to allow adequate nutrition by an enteral route.**
 - a) In your opinion, in an infant *with* an intact ileocecal valve, how short must the gut be to be inadequate: _____ cm.
 - b) In your opinion, in an infant *without* an intact ileocecal valve, how short must the gut be to be inadequate: _____ cm.
- 2) **How many cases of short gut syndrome meeting one of these definitions are seen at your institution per year?**
 - A) 0–2 B) 3–5 C) 6–8 D) 9–11 E) > 11
- 3) **Do neonatologists at your institution discuss treatment options when talking with parents about short gut syndrome?**
 - Yes No Unknown
- 4) **Who at your institution is the primary discussant regarding treatment options?**
 - A) Neonatologist
 - B) Gastroenterologist
 - C) Pediatric Surgeon
 - D) Unknown
- 5) **Which treatment options are usually discussed with parents at your institution?**
 - A) Enteral feeding only; no long term total parenteral nutrition (TPN)
 - B) IV fluids as major source of nutrition; no TPN
 - C) Long term TPN only
 - D) Transplant
 - E) Palliative surgery (i.e., gut lengthening surgery)
 - F) unknown
- 6) **Which treatment option is most often recommended to parents?**
 - A) Enteral feeding only; no long term total parenteral nutrition (TPN)
 - B) IV fluids as major source of nutrition; no TPN
 - C) Long term TPN only
 - D) Transplant
 - E) Palliative surgery (i.e., gut lengthening surgery)
 - F) unknown
- 7) **Which of the following treatment options are currently practiced at your institution?**
 - A) Enteral feeding only; no long term total parenteral nutrition (TPN)
 - B) IV fluids as major source of nutrition; no TPN
 - C) Long term TPN only
 - D) Transplant
 - E) Palliative surgery (i.e., gut lengthening surgery)
 - F) unknown
- 8) **What is your impression of the 3-year survival following long term TPN only.**

0%	20%	40%	60%	80%	100%
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- 9) **What is your impression of the 3-year survival following palliative surgery repair.**

0%	20%	40%	60%	80%	100%
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- 10) **What is your impression of the 3-year survival following transplantation.**

0%	20%	40%	60%	80%	100%
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- 11) **What is your impression of the quality of life following long term TPN only.**

Normal	Mildly restricted	Moderately restricted	Severely restricted
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- 12) **What is your impression of the quality of life following palliative surgery repair.**

Normal	Mildly restricted	Moderately restricted	Severely restricted
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- 13) **What is your impression of the quality of life following transplantation**

Normal	Mildly restricted	Moderately restricted	Severely restricted
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- 14) **Are your impressions primarily derived from the experience of your institution or by publications.**

Experience	Publication
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Fig 1. Short gut questionnaire.

both with and without an intact ileocecal valve (Table 1).

Both the neonatologists and pediatric gastroenterologists agreed that the use of TPN was discussed nearly always with the parents (>95%) and recommended nearly always (>90%). They disagreed, however, about the discussion and recommendation of alternative treatment options. Pediatric gastroenterologists were far more willing to discuss and recommend a less aggressive treatment option (enteral feeding only) and tended to discuss and recommend more aggressive treatment options (transplant and gut-lengthening surgery; Table 1).

Although both groups agreed that TPN was discussed and recommended nearly always, they sharply disagreed about the outcomes of its use. Neonatologists were significantly more pessimistic about 3-year survival with the use of TPN (Table 1). They were also far more pessimistic about the quality of life of patients on TPN. Two thirds of the respondents from both specialty groups viewed quality of life on TPN as moderately restricted, but the remaining neonatologists primarily viewed quality of life as severely restricted, whereas the remaining pediatric gastroenterologists primarily viewed quality of life as mildly restricted.

- 1) How many cases of hypoplastic left heart syndrome are seen at your institution per year?
A) 0–2 B) 3–5 C) 6–8 D) 9–11 E) > 11
 - 2) Do neonatologists at your institution discuss treatment options when talking with parents about hypoplastic left heart syndrome?
Yes No Unknown
 - 3) Who at your institution is the primary discussant regarding treatment options?
A) Neonatologist
B) Cardiologist
C) Cardiovascular Surgeon
D) Unknown
 - 4) Which treatment options are usually discussed with parents at your institution?
A) Comfort care
B) Transplant
C) Palliative surgery (i.e., Norwood/Fontan repair)
D) unknown
 - 5) Which treatment option is most often recommended to parents?
A) Comfort care
B) Transplant
C) Palliative surgery (i.e., Norwood/Fontan repair)
D) Unknown
 - 6) Which of the following treatment options are *currently* available at your institution?
A) Transplant
B) Palliative surgery (i.e., Norwood/Fontan repair)
C) Both
D) Neither
 - 7) Is the option of immediate palliative surgery followed by planned transplantation when a heart becomes available... (circle all that are applicable)
A) Discussed with parents at your institution?
B) Recommended at your institution?
C) Neither
D) Unknown
 - 8) What is your *impression* of the 1-year survival for palliative surgery repair.
0% 20% 40% 60% 80% 100%
 - 9) What is your *impression* of the 1-year survival for transplantation.
0% 20% 40% 60% 80% 100%
 - 10) What is your *impression* of the quality of life following palliative surgery repair.
Normal Mildly restricted Moderately restricted Severely restricted
 - 11) What is your *impression* of the quality of life following transplantation
Normal Mildly restricted Moderately restricted Severely restricted
- Are your impressions primarily derived the experience of your institution or by publications.
Experience Publication

Fig 2. Hypoplastic left heart syndrome questionnaire.

There were few differences in the perception of surgical outcome between the neonatologists and pediatric gastroenterologists (Table 1). The only difference was that neonatologists perceived the quality of life after palliation as worse than did pediatric gastroenterologists. Again, although approximately two thirds of the respondents from both specialty groups viewed quality of life after palliative surgery as moderately restricted, nearly half of the remaining neonatologists believed that quality of life was severely restricted, whereas none of the remaining pediatric gastroenterologists viewed it that way.

Table 1 also presents similar data from the HLHS survey of neonatologists and cardiologists. Neonatologists and pediatric cardiologists strongly disagreed about the role of neonatologists in discussion with families. They agreed that comfort care only is not discussed usually and that all surgical treatment options are discussed nearly always. The majority of both groups recommend Norwood surgery; only a minority recommend other treatment.

There are very significant differences in the perceptions of the median quality of life after the surgical options; the cardiologists are more optimistic than are the neonatologists. The disagreements about 3-year survival after surgery are much less pronounced.

There were 11 points of cross-disease comparison of interprofessional differences. There were sharp differences in the extent of interprofessional difference between the SBS respondents and the HLHS respondents in only 2 of the 11 points. First, neonatologists were less likely than were pediatric gastroenterologists to recommend transplant for SBS ($P = .046$), but they were as likely as the pediatric cardiologists to recommend heart transplant for HLHS ($P = .3$). Second, both neonatologists and pediatric gastroenterologists had similar perceptions of the quality of life after transplantation ($P = .86$), but neonatologists were much more pessimistic than were pediatric cardiologists about the quality of life after transplantation ($P = .001$).

TABLE 1. Interprofessional Differences

	Short Gut			HLHS		
	Neonatologists	Pediatric Gastroenterologists	<i>P</i>	Neonatologists	Pediatric Cardiologists	<i>P</i>
Neonatologist discusses options	39/47 (83%)	3/26 (12%)	<.001	56/72 (78%)	19/38 (50%)	.005
Length of bowel inadequate for enteral nutrition with an intact ileocecal valve	20 (8.0) cm* (10–50)†	31 (20) cm* (10–100)†	<.001	N/A	N/A	N/A
Length of bowel inadequate for enteral nutrition without an intact ileocecal valve	34 (10) cm* (15–50)†	52 (25) cm* (15–125)†	<.001	N/A	N/A	N/A
Discuss less aggressive treatment options	15/68 (22%)	18/38 (47%)	.013	8/71 (11%)	11/42 (26%)	.066
Discuss palliative surgery	27/68 (40%)	22/38 (58%)	.11	71/71 (100%)	41/42 (98%)	.37
Discuss transplant	29/68 (43%)	24/38 (63%)	.068	63/71 (89%)	38/42 (90%)	1.0
Recommend less aggressive treatment options	5/65 (8%)	8/38 (21%)	.08	18/64 (28%)	2/39 (5%)	.004
Recommend palliative surgery	3/65 (5%)	8/38 (21%)	.02	40/64 (62%)	33/39 (85%)	.024
Recommend transplant	9/67 (13%)	8/38 (21%)	.046	10/64 (16%)	10/39 (26%)	.30
Perceived median 3-year survival—TPN	60%	80%	.001	N/A	N/A	N/A
Perceived median 3-year survival—palliative surgery	60%	60%	.35	60%	60%	.257
Perceived median 3-year survival—transplant	40%	40%	.15	80%	80%	.097
Perceived median quality-of-life—TPN	Moderately restricted	Moderately restricted	.011	N/A	N/A	N/A
Perceived median quality-of-life—palliative surgery	Moderately restricted	Moderately restricted	.019	Moderately restricted	Mildly restricted	<.001
Perceived median quality-of-life—transplant	Moderately restricted	Moderately restricted	.86	Moderately restricted	Mildly restricted	.001

The number and mean (unless otherwise indicated) of the responses, by specialist, to each question.

* Mean ± standard deviation; †, range.

Intrainstitutional Dissonance

Having presented the data about differences in what is discussed, recommended, and perceived among specialty groups in general, we now turn to data about differing opinions (dissonance) among specialists at the same institution. This dissonance about whether to discuss or recommend an option and about whether an option is available at that institution is particularly troubling because of the resulting potential for conflicting counseling of parents.

Table 2 presents the data about intrainstitutional dissonance among specialists at the same institution. For example, the first line shows that in 10 of the 26 institutions (38%) the neonatologists and pediatric gastroenterologists disagreed as to whether palliative surgery (gut lengthening surgery) was discussed with the parents. But at only 1 of the 33 institutions (3%) did neonatologists and pediatric cardiologists disagree as to whether palliative surgery (Norwood/Fontan repair) was discussed with the parents. This represents a 13 times greater relative risk for dissonance in the discussion of SBS palliative and HLHS palliative options.

Overall, there was considerable dissonance about each of these issues between neonatologists and pediatric gastroenterologists at the same institution. For any given item listed in Table 2, the percent of institutions with dissonance ranged from 16% to 50% with a mean of 34% dissonance among specialists at the same institution. There was less dissonance between the neonatologists and the pediatric cardiologists at the same institution ranging from 3% to 27% of institutions with dissonance on any given item with a mean of 14%. It is particularly surprising that the highest degree of dissonance is about what is

discussed and locally available and that there is much less dissonance about what is recommended.

Consonant institutions are those in which there is no dissonance. Only 1 of the 25 institutions for which we have a full set of responses from both specialists was totally consonant on all the items related to SBS. In contrast, 7 of the 28 institutions for which we have a full set of responses from both specialists were consonant on all the items related to HLHS (*P* = .080). No institution was dissonant for more than four items on the table for either disease.

DISCUSSION

Our data reveal that there are substantial disagreements in treatment options discussed and recommended by neonatologists, pediatric gastroenterologists, and pediatric cardiologists. As Table 1 demonstrates, neonatologists, pediatric gastroenterologists, and pediatric cardiologists, as professionals, disagree about many aspects of management strategies and outcome. As Table 2 demonstrates, these professional differences result in considerable dissonance among the professionals at the same institution. These disagreements are present for both SBS and HLHS, but they seem to result in somewhat less intrainstitutional dissonance in the case of HLHS.

Some of these disagreements seem paradoxical. It is not immediately apparent why there is intrainstitutional dissonance over what is available at a given institution. Shouldn't the heads of the two involved programs at a given institution know what therapies are available at their institution? It also is not immediately apparent why there are interprofessional differences about recommending alternatives to TPN in the management of SBS. If the pediatric gastroenterologists (who are more optimistic about long-term

TABLE 2. Intrainstitutional Dissonance

	Neonatologists Versus Gastroenterologists 46 Potential Pairs	Neonatologists Versus Cardiologists 46 Potential Pairs	Relative Risk (95% Confidence Interval), P
Option is discussed			
Palliation	10/26 (38%)	1/33 (3%)	13 (1.7–93), P = .0017
Transplant	11/26 (42%)	6/33 (18%)	2.3 (0.99–5.4), P = .082
Comfort care	13/26 (50%)	9/33 (27%)	1.8 (0.93–3.6), P = .13
Option is recommended			
Palliation	4/25 (16%)	4/28 (14%)	1.1 (0.31–4.0), P = .83
Transplant	5/25 (20%)	4/28 (14%)	1.4 (0.42–4.6), P = .85
Comfort care	8/25 (32%)	4/28 (14%)	2.2 (0.77–6.5), P = .22
Option is locally available			
Palliation	11/27 (41%)	3/33 (9.1%)	4.5 (1.4–14), P = .010
Transplant	11/27 (41%)	4/33 (12%)	3.4 (1.2–9.4), P = .025

The number (and percent) of dissonant responses for each question within paired institutions where there are both neonatology and either gastroenterology or cardiology specialty training programs.

TPN outcome) are willing to consider both more and less aggressive alternatives, why aren't the neonatologists (who are more pessimistic) willing to consider alternatives?

One speculation, the dissemination explanation, is that the first of these paradoxes is a function of the relevant newness of the alternatives to TPN. Because these therapies are just becoming available at many institutions, there may have been less time for widespread appreciation of local availability of these procedures. Alternatively, the timing explanation asserts that this dissonance relates to the fact the surgical alternatives to TPN are implemented rarely in the immediate neonatal period; they are more likely to be discussed, recommended, and chosen after problems with TPN emerge and/or it becomes apparent that enteral feeding will not be successful. At that later point, the neonatologists are usually no longer active participants in the management. This nonparticipation at a crucial time may result in differences in their awareness of what is locally available from the pediatric gastroenterologists who continue as active participants. Supporting both of these alternatives is the fact that there is much less intrainstitutional dissonance about the local availability of treatment options for HLHS. Those treatment options were introduced years earlier with comparatively extensive outcome data currently available^{15–20} and must be chosen during the neonatal period.

Timing seems to be the central factor in explaining the unwillingness of the neonatologists to consider both less aggressive and more aggressive alternatives, despite their pessimism about the outcome on TPN. Because decisions about alternatives usually are faced at a later time when the neonatologists are no longer involved, it is relatively easy for them to consider TPN, which works in the short run and enables them to send the baby home. Dissemination of information is not entirely irrelevant. If and when data emerge that indicate that the outcome of these procedures is better than TPN, timing problems will be overshadowed. Neonatologists then may begin to discuss and recommend alternatives (for now or in the future) while they are involved actively in the management of the baby.

This study is limited by problems intrinsic with all surveys: we can record only what the responding

program directors said that they do and cannot measure what they actually do. We also are limited by the fact that we are asking them about the behavior of their service as a whole and not just their own behavior. However, the rate of response, which is often a problem for survey research,²¹ is not a problem in this study with an overall response rate of 79%. Although the sample size itself is small, the group that we are sampling is also small; this type of response rate means that our estimates do reflect accurately the opinions of the entire group of program heads.

Two major ethical issues arise from our data. The first is the question of how much data a clinician needs about new therapeutic interventions before beginning to discuss and recommend them. The tremendous variance in all the relevant professions shows that there is no accepted professional standard to which individuals can appeal. As we have shown in our earlier research, a standard for discussing and recommending alternatives not yet adequately supported by data has not yet emerged in either the legal or the ethical literature.¹ The second ethical issue is the potential for much confusion of parents at a difficult time when the counseling physicians at the same institution discuss and recommend different options. Although there is no need to enforce unanimity on professionals who honestly disagree, there is also no need to confuse parents needlessly if these disagreements simply reflect the failure of the professionals within an institution to educate each other about their alternative views and practices.

A standard for discussing and recommending new alternatives clearly is needed. The requirement of informed consent includes the discussion of alternatives, but surely does not require discussion of newly conjectured, unsupported-by-data alternatives. At what point do these alternatives become sufficiently plausible, so that the parents are entitled to a discussion of them as alternative? Even if difficult to develop, the needed professional standard would answer that question.

The problem of the intrainstitutional dissonance is easier, in theory, to solve. Colleagues as individuals and subspecialty services as groups should talk to each other before individual discussions

with families with the goal of ensuring that there is a clear understanding of differing beliefs. If those discussions reveal true disagreements, they can be presented reasonably to families in a collegial fashion. But these disagreements may disappear after an open and frank discussion among professionals. An alternative solution would be for neonatologists to refuse to discuss these issues with the families and refer them to pediatric gastroenterologists who will deal with these issues at the latter stage when they arise. We believe that families who are first confronting the problem of a baby with SBS would object if the managing neonatologist refused to discuss these issues. Therefore, we support our first recommendation of dialogue among professional groups both in general and in individual institutions.

Neither the recommendation of the development of standards nor the recommendation of interprofessional dialogue will be easy to implement. However, our data show that these implementations are needed to deal with real problems existing currently.

REFERENCES

1. Cooper TR, Caplan WD, Garcia-Prats JA, Brody BA. The interrelationship of ethical issues in the transition from old paradigms to new technologies. *J Clin Ethics*. 1996;7:243-250
2. Collins JB, Georgeson KE, Vicente Y, et al. Short bowel syndrome. *Semin Pediatr Surg*. 1995;4:60-72
3. Figueroa-Colon R, Harris PR, Birdsong E, et al. Impact of intestinal lengthening on the nutritional outcome for children with short bowel syndrome. *J Pediatr Surg*. 1996;31:912-916
4. Gambarara M, Ferretti F, Bagolan P, et al. Indications for small bowel transplantation in children with gastrointestinal pathology on long-term parenteral nutrition. *Transplant Proc*. 1996;28:2706-2707
5. Baliga P, Reuben A, Baker R, et al. Intestinal transplantation: an early experience. *Transplant Proc*. 1996;28:2734-2735
6. Beath SV, Kelly DA, Buckels JA, Mayer AD. Initial experience of pediatric combined small bowel and liver transplantation. *Transplant Proc*. 1996;28:2736-2737
7. Bianchi A. Autologous gastrointestinal reconstruction. *Semin Pediatr Surg*. 1995;4:54-59
8. Flageole H, Adolph VR, Sigalet DL, et al. The Bianchi procedure in a patient with jejunal atresia. *Eur J Pediatr Surg*. 1997;7:361-363
9. Bianchi A. Longitudinal intestinal lengthening and tailoring: results in 20 children. *J R Soc Med*. 1997;90:429-432
10. Weber TR, Powel MA. Early improvement in intestinal function after isoperistaltic bowel lengthening. *J Pediatr Surg*. 1996;31:61-63
11. Langnas AN, Shaw BW, Antonson DL, et al. Preliminary experience with intestinal transplantation in infants and children. *Pediatrics*. 1996;97:443-448
12. Norwood WI, Kirklin JK, Sanders SP. Hypoplastic left heart syndrome: experience with palliative surgery. *Am J Cardiol*. 1980;45:87-91
13. Bailey LL, Concepcion W, Shattuck H, Huang L. Method of heart transplantation for treatment of hypoplastic left heart syndrome. *J Thorac Cardiovasc Surg*. 1994;107:934-940
14. Caplan WE, Cooper TR, Garcia-Prats JA, Brody BA. Diffusion of the innovative approaches to managing hypoplastic left heart syndrome. *Arch Pediatr Adolesc Med*. 1996;150:487-490
15. O'Kelly SW, Bove EL. Hypoplastic left heart syndrome: terminal care is not the only option. *Br Med J*. 1997;314:87-88
16. Allan LD, Apfel HD, Printz BF. Outcome after prenatal diagnosis of the hypoplastic left syndrome. *Heart*. 1998;79:371-373
17. Hehrlein FW, Yamamoto T, Orime Y, Bauer J. Hypoplastic left heart syndrome: which is the best operative strategy? *Ann Thorac Cardiovasc Surg*. 1998;4:125-132
18. Bove EL. Current status of staged reconstruction for hypoplastic left heart syndrome. *Pediatr Cardiol*. 1998;19:308-315
19. Kern JH, Hayes CJ, Michler RE, et al. Survival and risk factor analysis for the Norwood procedure for hypoplastic left heart syndrome. *Am J Cardiol*. 1997;80:170-174
20. Dapper F, Bauer J, Kroll J, et al. Clinical experience with heart transplantation in infants. *Eur J Cardiothorac Surg*. 1998;14:1-6
21. Asch DA, Jedrzewski MK, Christakis NA. Response rates to mail surveys published in medical journals. *J Clin Epidemiol*. 1997;50:1129-1136

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