NEGOTIATING PKU:
INSIGHTS FROM NEW ZEALAND

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ABSTRACT

This paper explores the experience for nine New Zealanders of living with Phenylketonuria (PKU). PKU is an inherited disorder of metabolism, detected by newborn screening and treated primarily through adherence to a strict and rather unpalatable diet which current medical opinion – suggests should be lifelong. Our goal is to problematise this biomedical notion of 'adherence' to such a diet, and to do so by drawing on two areas of expert knowledge which are relatively unexplored in the medical literature – the views of people living with the condition and the social science literature on food and consumption. Both of these systems of knowledge provide useful contextual information to explore the complexity of adherence and move it beyond a simple notion of rational decision making. We suggest that biomedical 'problems' such as adherence are quite usefully addressed by hybrid research plans such as the one which underpins this project. However conveying the resulting information back outwards into the biomedical sphere is rather more difficult for it implies the existence of a community of hybrid readers and a hybrid literature in which to place such work.

'At the simplest, biological level, by the act of eating and absorption of food, we become what we eat' Lupton (1996:16).

INTRODUCTION

This research explores the experience of living with Phenylketonuria (PKU) for a group of nine New Zealand adults. While this condition has the advantage of being a treatable genetic disorder (through modification of the diet as explained below), the treatment itself is arduous, and compliance with medical treatment creates significant social costs to the individuals concerned. As such, the situation invites anthropological study, because understanding how people manage the challenges of living under such circumstances can provide useful insights for the medical domain.

The focus of this paper then, is to review the medical literature on the dietary management of PKU which currently explains the medical problem of managing the PKU diet through the concept of 'adherence'. We then attempt to translate some of the social science literature on food consumption across onto this literature to both problematise the concept of adherence and to theorise the missing social context to food consumption currently evidenced in the medical literature. We suggest that by synthesising these two broad fields of knowledge it may be possible to better understand the worlds of people living with PKU. Finally we present ethnographic data gathered from people affected by PKU on the challenges of living with recommendations for such intensive dietary management. In their interviews, these people explained how such treatment has widespread consequences, touching on all spheres of life (delineated in this article as the medical, social, and personal spheres). The end result of which is that they are forced to become expert negotiators of the demands both within and between these spheres of living. The many different factors which the interviewees suggest are being weighed in their decision to adhere or not to adhere to the PKU diet provide a richer, more contextual view of the issue of treatment adherence which could aid medical practitioners in their interactions with PKU adults.

Our project should then be understood as a small contribution to the emerging New Zealand work which investigates the social context of living with a variety of genetic conditions and addresses an explicitly hybrid research literature informed by both medical and social science knowledges to do so. In this emerging field, Julie Park's long ranging New Zealand study on the experience of living with haemophilia is an exemplary methodological signpost for its sustained attention to the complex interplay between treatment adherence, biomedical implications of non-adherence, and the experience of gendered ethnicity and family life (Park and Strookappe 1996; Park 1998; Park, Scott and Benseman 1999; Park 2000; Park 2005).

THE MEDICAL BASIS FOR A SOCIAL PROBLEM

PKU is an inherited disorder of amino acid metabolism with an incidence of approximately one in 15,000 births of European descent, in which the body is unable to convert the amino acid phenylalanine to another amino acid, tyrosine, due to mutations in the gene coding for the enzyme phenylalanine hydrolase. (Amino acids are the building blocks of proteins, which perform a wide variety of functions in the cell and are the major structural components of all body tissue, and are also involved in a wide range of metabolic processes). To date there are over 328 mutations identified both in the gene for this enzyme and the immediate regions specifically adjacent to the gene (flanking regions). As a result of this error in metabolism, phenylalanine accumulates in the blood and other tissues, leading to severe mental retardation in the untreated individual. Although the precise mechanism for the cause of the mental retardation and related clinical conditions are still not fully understood, there is strong evidence that high levels of phenylalanine in the brain will interfere with normal neurotransmitter function. Fortunately, newborn testing, through the heel prick or 'Guthrie' Test, which is carried out immediately after birth, has been available for PKU for approximately the past forty years. Significantly, today's adults with PKU therefore comprise the first generation of adults who have been diagnosed with (and ideally treated for) PKU since infancy. Historically, such people were frequently undiagnosed, and therefore untreated, and lived out their lives confined to mental institutions.

When PKU is detected in infancy, and dietary treatment is begun within the first few days of life, normal development is possible. However medical knowledge is no less dynamic and mutable in its construction and its transmission than any other field of knowledge, and while it was once thought that discontinuation of the therapeutic diet was acceptable beyond adolescence with no adverse consequences, more recent research shows that discontinuation of the PKU diet, even in adulthood, could have serious negative neurological effects. For this reason, lifelong adherence to the PKU diet is now suggested, and many New Zealanders living with PKU have recently grappled with the conflict of deciding whether or not to continue (or even return to) their childhood diet. As the treatment recommended for individuals with PKU is quite rigorous, even for those who willingly embrace the programme, the rate of adherence to this dietary regimen declines as individuals with PKU age.

The orthodox treatment for PKU consists of a restricted intake of foods high in phenylalanine, consumption of a specially prepared amino acid mixture (or protein substitute), a highly regulated intake of certain foods containing moderate amounts of phenylalanine combined with the use of medical foods (and some natural foods) very low in phenylalanine, and frequent blood tests to monitor phenylalanine levels. Clearly, the treatment requires an extraordinary degree of self-surveillance and impacts hugely on the social act of food consumption.

Phenylalanine is found in most natural sources of protein, and so high-protein foods, such as meat, poultry, fish, eggs, cheese, milk products, nuts, most flour products (such as bread and cake), beans, lentils, and meat substitutes are either eliminated from, or greatly reduced in, the PKU diet. Often, natural protein intake is reduced to less than six grams per day ('PDPHD' 1993). Even so, phenylalanine cannot be entirely excluded from the diet. Thus, low-phenylalanine foods (e.g., potatoes, corn, peas, and cereal products) are allowed in controlled amounts. An exchange system, 'whereby one food can be swapped for another of an equivalent phenylalanine content' (MacDonald 2000: S139), is often used as a practical tool for regulating the PKU diet. The number of permissible daily exchanges is individually specific and is calculated by a metabolic physician or a dietician. The design of the exchange system also varies internationally, with New Zealand using a less finely calibrated system involving fifty milligram exchanges (see Figure 1). Most patients with classical PKU are allowed between four and eight (fifty milligram) phenylalanine exchanges daily (MacDonald 2000), however, the participants in this project seemed to be allowed slightly more. Excluding times of pregnancy or medical trials (when allowances may be more restricted), their permitted daily exchanges ranged between eight and twenty milligrams per day.

TRANSLATING BETWEEN THE SOCIAL SCIENCE AND MEDICAL LITERATURE

After reading the preceding section on the medical explanation of PKU and its associated dietary treatment regime, that (along with the consequences of not adhering to the diet) would seem sufficiently sobering motivating factors to adhere to the PKU diet (for those who do not live with PKU). However, the subsequent interviews with people living with PKU will reveal instead a situation of constant tensions, distractions and practical difficulties which thoughtful people who live with PKU negotiate daily in their decisions concerning how to live with their condition. In some cases the considered outcome of these negotiations is to *not* adhere to the diet. However, to see the difficulties of following the diet solely as a medical problem of dietary 'non-compliance', neglects the social dimensions of food consumption – an area in which people living with PKU and anthropologists of food are experts. Regrettably, the nutritional or sociobiological perspective, evident in so much of the medical literature on PKU, tends to take an instrumental view on food and eating, understanding food habits and preferences in relation to the physiological operation of the body, thus minimising the social aspects of dietary adherence and the experience of pleasure in eating. The result has been the development of some mechanistic models of health behaviour, which address only the psychologically individualized aspects of health-related choices in food consumption minimising or ignoring altogether the social context and meaning of eating.

Type of Food	*Quantity of Food = One 50mg Exchange*
Egg (chicken)	⅙ egg
Cottage cheese	½ Tablespoon
Processed cheese	¼ slice
Parmesan cheese	⅓ Tablespoon
Cheddar cheese	4.4 g
Pork, bacon (lean, cooked)	3 g
Chicken (cooked)	4.3 g
Beef (roast, lean)	3.1 g
Export Gold, Tui, D.B. Drought beer	1 can
Saltine cracker	¾ cracker
Commercial white/wheat bread	½ medium slice
Kelloggs Cornflakes	½ cup
Kelloggs Sultana Bran (Raisin Bran)	3 ½ Tablespoons
Brown/white rice (boiled)	3 Tablespoons
Spaghetti (dry, uncooked)	20 g
McDonalds fries	⅓ small serving
KFC coleslaw	1 small tub (110 g)
Ice cream, vanilla	3 Tablespoons
Yogurt, plain and flavoured	1 Tablespoon
Peanut butter	½ teaspoon
Almonds	3 nuts
Peanuts	11 nuts
Potato chips (crisps), all flavours	½ cup (6 or 7 small chips)
Cadbury dairy milk chocolate bar	2 cubes
Baked beans (canned)	1 ½ Tablespoons
Potato (roast)	1 small egg sized potato
Mixed vegetables, frozen, boiled, drained	¼ cup
Corn (sweet, canned)	3 Tablespoons
Popcorn	3 cups
Pure fruit juices	*Free food*
Black tea, black coffee	*Free food*
Mushrooms	*Not more than 3 small daily*
Butter or Margarine	*Free food*
Carrots	*Free food*
Broccoli	*Free food*
Lettuce	*Free food*
Tomato	*Free food*
Popsicles or ice blocs (water or fruit juice)	*Free food*
Sugar (white, brown, castor, icing, raw)	*Free food*
Oranges	*Free food*
Strawberries	*Free food*
Bananas	*Not more than 1 daily*
Apples	*Free food*

Figure 1. Chart of 50 mg Exchanges.
Data taken from Nutrition Services: Auckland Hospital, 2005

The first example of these models is the Health Belief Model, which centres on the assertion that threat perception is a primary motivating factor leading to positive health behaviour. The perception of threat includes the elements of personal susceptibility to the condition and the perceived severity of the threat. Relevant to this would be the complaints the primary author received from participants of sleepiness, irritability and lack of manual dexterity when 'not adhering to the diet' or the manner in which many of them viewed dietary adherence as an 'insurance policy' for the future. Additionally, for behaviour to be influenced towards adherence, it must be believed that there is an effective action that can be initiated to reduce this threat (a belief currently undermined by the changing medical opinions of the values of adhering to the PKU diet). Finally, for behaviour to become health enhancing, the barriers or costs involved in implementing this action (such as the cost of protein supplements and the time consuming nature of PKU recipes) must be viewed to be outweighed by the benefits incurred. It has also been suggested that a 'cue' or 'trigger' must be present to stimulate the implementation of the health-related behaviour (Abraham and Sheeran 2000; Quine, Rutter and Arnold 2000; Gochman 1988). We agree that elements of this model arose in interviews but also suggest that previous criticisms of the model, such as its lack of consideration of the value of habit in eating behaviours and its failure to distinguish between medication and the more social aspects of adherence to treatment protocols, still apply.

The Theory of Planned Behaviour is another cognitive model often used in biomedical literature to explain health behaviours. It posits that behavioural intention is the single most important factor in predicting an individual's behaviour. This is directly influenced by overall feelings of favourableness or unfavourableness toward an action and the individual's perception of what others would like for them to do (Carter 1991; Gochman 1988; Quine, Rutter and Arnold 2000). This theory has been applied in the context of dietary adherence of people with PKU in the USA (Finkelson, Bailey and Waisbren 2001: 515) suggesting that the strength of support from one's social network influences decisions to return to the PKU diet, as would belief in the diet's efficacy and the individual's perception of the diet's manageability. The members of Finkelson, Bailey and Waisbren's study who did not maintain the diet revealed an inability to avoid temptation when eating in restaurants or at social functions, difficulties in paying for the medical foods and protein substitute, and forgetting to take the protein substitute every day (Finkelson, Bailey and Waisbren 2001: 515). These barriers were also relevant to the people in this study. However the strongly rational cast to this model for explaining health behaviours, as noted by Gibbons, Gerrard and Ouelette et al (2000: 137), fails to explore those behaviours which are 'less rational, but also exciting or enticing' other than in the negative sense of a constraint on purposeful action. The positive pull of such sensations is better described in the social science literature on food which we will explore further on in this section.

Attribution Theory is yet another health behaviour model which has been used in the biomedical literature. Antshel, Brewster and Waisbren (2004) have applied it directly to the experience of people with PKU to explain treatment adherence (or lack thereof) for PKU sufferers in the USA. This model, which was developed over time (from the late 1950s to the late 1960s) from the theories of social psychologists Fritz Heider, Edward Jones, Keith Davis and Harold Kelley, explores the ways in which people attribute certain causes to events in their lives and describes how the subsequent behaviours and emotions are intimately and logically connected to these decisions about the nature of the cause. Attribution is a complex phenomenon consisting of several different elements, which are all considered to affect an individual's response to their illness and can predict their ability to cope. These include whether the locus of causation is internal or external to the person, the extent to which the causes are believed to be controllable or uncontrollable, the extent to which a person expects further situations to have similar outcomes, and also whether a cause has a limited or wide variety of outcomes (Lewis and Daltroy 1991: 95). A perceived internal locus of control has been linked to adherence to many different health-promoting behaviours, including, as Antshel, Brewster and Waisbren (2004: 627) have noted, adherence to and belief in the PKU diet for affected children and their parents. Certainly our own study signalled a very high sense of personal responsibility and control over dietary adherence exemplified for many in the following statement: 'I'm on my own…. It's just me and the supermarket trolley' (8 October 2005, interview). However, while such statements are suggestive of a strong internally placed locus of control, participants also suggested a degree of emotional ambivalence over this responsibility – a point to which we will return.

There are many more models of health related behaviour in existence than those already cited (see Frank 2006 for a wider discussion), however this selection gives a flavour of the biomedically favoured explanations of health behaviour around the issue of adherence to the PKU diet. One limitation common to all cognitive models, or theories relating to rational or volitional choice, concerns the role of habit in adherence to medical regimens and preventive health behaviours. This deserves mention, as it appears from our own study that routine may play a considerable role for some people with PKU in the maintenance of their diet. In fact, one participant in this project even cited 'habit' as his primary motivation for adhering to the PKU diet (19 July 2005, interview). And although these models, and others like them, do raise many plausible factors relating to treatment adherence, adherence itself can be a problematic concept with a tendency to oversimplify reality. For example, Trostle has observed that treatment adherence, rather than simply being a cognitive decision to improve our health behaviours or not, 'can be shown to fulfil many other motivations at once, ranging from a desire to satisfy a physician or family members or peers to a work-related need or a desire to enhance or diminish specific side effects' (Trostle 2000: 52).

The social sciences approach, of course, tends to emphasise these broader contextual framings for apparently simple decisions by individuals in regard to their health behaviours. Given its obvious relevance, it is thus somewhat disturbing to observe that so little theoretical material from the social sciences tends to find its way across into a medical readership. The next few paragraphs set out some of the more obvious contributions to understanding 'adherence' which the sociology and anthropology of food can contribute.

For instance, the social science literature on food recognizes that while the practices surrounding the consumption of food '…may be governed by biological needs and the availability of foodstuffs in the first instance…, these practices are then elaborated according to cultural mores. Food practices are therefore far more complex than a simple nutritional or biological perspective would allow' (Lupton 1996: 7). People living with PKU understand first hand that food and eating habits and preferences are not simply matters of biological necessity, but incorporate any number of subjective meanings, serve myriad social functions, and are central to human understandings and experiences. As one interviewee noted, '…everything in life revolves around food. It's really hard to get away from it' (8 October 2005, interview).

Food is highly symbolic, and for that reason, is a powerful communicator. It can communicate (and even elicit) emotion, celebration, or even personal attributes of the consumer, such as self-control. Eating behaviours thus also help to form individual identity. The act of eating transfers not only nutrients and sustenance to the consumer, but also the symbolic values and characteristics inherent in the food item: '…a widespread feature of human culture is the idea that the absorption of a given food, particularly when occurring repeatedly, can have the effect of transferring certain symbolic properties of that food into the very being of the eater' (Beardsworth and Keil 1997: 54). Although this idea dates back to ancient times, in the modern world, this concept is taken advantage of by advertising, which explicitly attaches positive values and images to food items (see Mintz 1996; Lupton 1996; Falk 1994). The ability of individuals with PKU to associate these positive commercial images with themselves is impaired, however, by their dietary restriction which forbids the consumption of many such foods. People with PKU may, therefore, instead identify themselves with the images accompanying the foods they *do* eat, and these associations may be much less positive.

The appearance of difference (or even strangeness or vulgarity) in food-related practices is also communicative, holding '… an extraordinary power in defining the boundaries between 'us' and 'them'' (Lupton 1996: 26), and 'mark[*ing*] off insiders from outsiders' (Peckham 1998: 172). In fact, throughout history, and in many different areas of life, '…there seems to be a continuous tradition of establishing group memberships through eating' (Back 1977: 32). Examples of this abound. For instance, the gender specific (Western) consumption of 'light' or 'heavy' foods, and serving size (e.g. Lupton 1996; Beardsworth and Keil 1997; Counihan 1999); the socioeconomic classes affinities for certain foods (Bourdieu 1984); and the creation and maintenance of ethnic identity through food-related practices (Peckham 1998: 172) all serve to generate group boundaries. In the context of PKU, food related practices establish one's membership in groups formed on the basis of health, or what Sontag refers to as the 'kingdom of the sick' (1978: 3). In addition to group formation through the expression of similarity and difference, group cohesion and community can also be created in the physical expression of sharing food, or eating together (Counihan and Van Esterik 1997: 3). However, the refusal to share food is equally meaningful. 'Because of the mandatory nature of food-sharing, food refusal and fasting have powerful social and symbolic weight' (Counihan and Van Esterik 1997: 3). In essence, because eating together implies intimacy (along with trust, friendship, and kinship) (Counihan 1999: 9, 13), refusal of food may indicate a rejection of that intimacy, and therefore serve as a 'sign of enmity and hostility' (Counihan 1999: 13). Most participants in this study have named the expectations involved with the sharing of food as one of the primary difficulties that they encounter in dietary adherence, and one of the most common reasons for the transgression of their diet.

Finally, a number of expectations are placed on eating habits and food-related values that reflect and shape the normative consumption patterns in today's society. These pressures are particularly significant for people with PKU who, due to their dietary restrictions and requirements, may be unable to meet many of these expectations. In addition to gendered expectations regarding eating and the social expectation to share food (mentioned above), is the pressure to conform. Mintz expounds upon to these 'powerful pressures toward sameness' (1996: 113). Back additionally stresses the lack of acceptance granted to deviation from the norm in this area, claiming, 'There is a general tendency to accord more respect toward differences in sexual patterns of other individuals and cultures than toward corresponding differences in eating habits' (Back 1977: 33). Contemporary Western society has also placed great value on the ideals of variety and the exotic in relation to dining experiences, forming a 'gourmet culture' among the economically privileged in Western societies, 'in which 'artistic', 'refined' and 'innovative' cuisine is valorised' (Lupton 1996: 145). As mentioned above, participants in this project have lamented the lack of variety that their PKU diet allows. This deficit must be felt particularly acutely in light of the high value society places on novelty in eating, and the positive symbols associated with 'sensation seeking' through the consumption of food.

This section reveals the ease with which the extant social science literature fills in the 'context' which Trostle (2000) notes is so important for understanding the complex decision making behind seemingly clear-cut decisions to adhere (or not) to the PKU diet. However, such rich theoretical context is not valuable without a simultaneous engagement with biomedical understandings of the significance of diet for the individual's health. It is to a possible synthesis of these knowledges as expressed in the reflections on the diet from those people primarily affected by it ie people living with PKU, that we now turn.

METHODS

Qualitative interviewing was chosen as the most appropriate and effective research technique to explore the lives and perspectives of these adults living with PKU as it is considered to be effective in establishing common themes or patterns between particular types of individuals (Warren, 2002: 85), '… unveil[*ing*] the distinctive meaning-making actions of interview participants' (*ibid*: 86), and uncovering motives and conflicts experienced by individuals. This research attempts to see PKU and treatment adherence through the eyes of an individual with PKU, thus giving us a glimpse into their world. 'The basic access to any social world is the accounts that people can give of their own actions and the actions of others. These accounts contain the concepts that the participants use to structure their world, and the 'theories' that they use to account for what goes on' (Blaikie, 1993: 176–177). Thus, the interviews conducted for this project sought to capture these accounts and tease from them the structuring concepts by which their experiences are shaped, and through which they are interpreted.

Therefore, nine extended open-ended interviews were conducted in 2005.

Following ethics approval for the research, obtained from the New Zealand Multi-Region Ethics Committee, the project participants were recruited via a third party (for reasons of patient confidentiality). Potential participants were located through a national database available to the National Metabolic Service, based in Auckland, New Zealand. In total, 46 letters describing the project and requesting the participation of interested parties were sent out to all PKU adults in the database over the age of eighteen. Eight people (and one mother of PKU children who had heard about the project through another participant) responded and agreed to participate.

The interviews lasted between 45 and 90 minutes (although no time limits were placed on the interviews), and several were followed up with further spoken or written communication – either initiated by the interviewer to further explore or confirm portions of the interview, or by the interviewee to bring up points that they forgot to mention during the interview, or to relate thoughts that they had had since that time. All interviews bar one were conducted in person, with the remaining interview being conducted over the telephone, as per the participant's wishes. Questioning was largely open ended and fluid, allowing the interviewee to help guide the discussion, and permissive of the emergence of new topics. The interviews usually began with general and impersonal questions such as, 'Please tell me a little bit about what PKU is, and how it's treated…', and usually the responses to even these first questions brought up topics that guided the following discussion. No script was used during the interviews, although the interviewer had in her mind a list of general areas to be touched on during the interview. These included: participant understandings of PKU, impressions of normalcy and difference in regard to PKU, identity formation, challenges of PKU and treatment adherence, coping mechanisms, change in experience over time, relationships with medical personnel and protocol, familial and social impact of PKU, emotional aspects of the experience of living with PKU, risk perception in relation to treatment adherence, personal agency and control, and the sensual aspects of the PKU diet. Some of these areas came to the researcher's attention through previous exposure to the medical and social science literature, and other topics came to light through an early analysis of the initial interviews, as will be explained further below.

The interviews were transcribed verbatim and then analysed using open, axial, and selective coding techniques, which are based in the broader disciplinary perspective of grounded theory (Strauss and Corbin 1998). The coding process involved a close reading of all data, labelling certain pieces of information according to the themes they depicted, and relating these themes to each other, forming a cohesive original argument that remains faithful to, or 'grounded' in, the data collected, while also contextualized within the broader background of relevant pre-existing social theories (discussed in detail below). Figure 2 depicts the schematic relationship between lower and higher order themes, which emerged from the coding process.

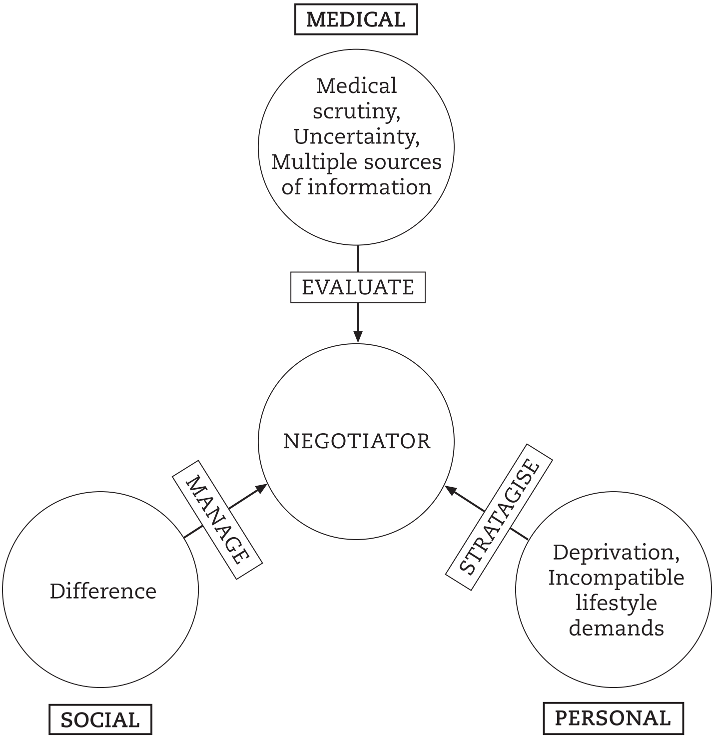

Figure 2. Diagram of the Organisation of the Research Results.
The three outer circles of this diagram represent the three spheres of life in which the participants in this project must negotiate their adherence to the PKU dietary recommendations: the medical sphere, the social sphere, and the personal sphere (as labelled). Within these circles are listed the structuring elements specific to that sphere.

It should be noted that true grounded theory research also involves a continuous interplay between the collection of data and its analysis, allowing for sensitivity to previous results in the alteration of the data collection method. Thus, grounded theory research should be cyclical in nature – collecting data, analysing data, altering the questions or methods as a result of the analysis, and then returning to the collection of further data. Although the scope of this research project was not large enough to permit an application of this principle on a grand scale, this aspect of grounded theory methodology was applied in a limited sense. The interviews for this project were collected in two different spans of time – the first in July of 2005, and the second in October of 2005. Although formal coding took place after all interviews had been collected, the first group of interviews were informally analysed, and interview questions and approaches were duly altered as a result of the initial findings. For example, the primary researcher was able to check the prevalence some of the early responses received by asking the second group of participants if they concurred with the earlier group's sentiments. Additionally, she found that the initial interviews brought to light the importance of topics that had not been previously considered – such as their experiences of risk, uncertainty, and medical experimentation – which were later incorporated into the interview format.

FROM THE MOUTHS OF THE 'EXPERTS': THE ETHNOGRAPHIC FINDINGS

The interviews revealed some remarkable trends and similarities in the world of people living with PKU in New Zealand. These common threads were the multiple structuring elements which are reacted to and worked around – or negotiated – in the incorporation of PKU into the participants' lives and which provide a type of skeleton to shape the participants' actions and life events. These elements were the concepts of medical scrutiny, uncertainty, multiple sources of information, difference, deprivation, and incompatible lifestyle demands. Each of these structuring elements presents certain challenges, which must be negotiated by the individual with PKU, and the core meaning of living with PKU identified by this project was the forced necessity to become a skilful negotiator in all of these areas. The method or aspect of negotiation employed in relation to each structuring element is dictated by the type of challenge it presents, which in turn is related to the sphere of life in which it occurs.

The interviewees in this project addressed challenges present in three different spheres of life: the medical, the social, and the personal. Clearly, one's life course is the point of intersection for all of these experiential elements. As Figure 2 illustrates, the challenges of the medical sphere are negotiated through evaluation of risk, various sources of (often conflicting) information, and the person's own priorities and values. The challenges of the social sphere are negotiated through the management of information and impressions, and ultimately the management of interpersonal relationships. The challenges of the personal sphere are negotiated through strategic incorporation of the PKU diet into one's life – a frequently incompatible and occasionally hostile environment. We will now discuss these three spheres and their negotiation in more detail.

MEDICAL SPHERE

One structuring element common to most accounts of living with PKU is the individual's perceived status as 'the object of medical scrutiny', or even 'guinea pigs'. This status is manifest in the constant and ongoing connection between the individual with PKU and the medical profession, with a continual monitoring of the individual's health and condition. Regular blood tests require continual contact with the medical world, as they must be mailed to a laboratory, and results must then be communicated back to the individual with PKU (as opposed to the somewhat more autonomous system for blood testing available to people with, for example, diabetes). Periodic appointments and communication with doctors, nurses, and dieticians likewise pepper the schedules of people with PKU. Many interviewees felt very acutely the 'newness' of their situation, and unlike people suffering from more established and understood conditions, they saw themselves as on the cutting edge of medical knowledge: '…when you're this age, you're sort of…learning with [*the medical community*], and…breaking ground with them' (19 July 2005, interview). This level of medical attention was seen by participants as both positive and negative. For instance, in general, the project's participants appeared to have a great respect and gratitude for medical advances, and frequently appreciated the heightened medical attention. At the same time, however, some resented their reliance on medical intervention, and many related past experiences of objectification: '…you sometimes feel like you are sitting in a gold fish bowl with everyone looking at you…all these people sort of focusing on you' (21 October 2005, interview).

Another structuring element emerging from interviewees' accounts of their lives with PKU was uncertainty. As the first generation of early-treated adults with PKU, many participants expressed in interviews the sentiment that no one, including the medical community, knows what the future holds for people with PKU. As one interviewee stated, '…if the oldest person in the world is only fifty that has got [*early-treated*] PKU, well, you know, you ask the question…are they going to live till they are seventy? Well, who knows? 'Cause the oldest person who has it is fifty' (20 October 2005, interview). Uncertainty was also felt by participants in relation to how long they would retain their mental faculties. In general, the people in this project viewed their situation as 'uncharted territory', or a 'no-man's-land'.

In negotiating this perceived deeply uncertain future, those who live with PKU must collect and evaluate information and impressions from many different sources. The still-developing knowledge of PKU and the resulting uncertainty mentioned above also leads to the administration of occasionally conflicting advice by the medical community. This competing information must also be evaluated by individuals with PKU. As previously noted, many participants expressed frustration at being told that they were able to come off the diet in their youth, only to be told to resume the diet later in adulthood. One person said:

> I was off it from when I was about ten or eleven until my early to mid twenties. Because there was a …PKU newsletter that mum used to read more than me – and there had been a conference, and they said in it that…the thinking then was diet for life, and going back – you know, you should be on it. And I went back to the paediatrician who…wanted to come off it, and he was like, oh, no, no, I haven't heard anything (18 October 2005, interview).

Many others have had similar experiences. At such an impasse, the onus is on the individual with PKU to make judgments as to what action would be in their best interests. Participants had also received conflicting advice as to what blood phenylalanine levels are acceptable or optimal. They expressed that '…every five years they change their theory about how things work and what you should and shouldn't be doing, and what's acceptable blood levels, and all this kind of thing' – 'you get told different things at different times' (8 October 2005, interview).

Medical expertise, however, is not the only source of information that must be evaluated by the PKU adult. In many instances, embodied and experiential knowledge makes the individual with PKU something of an expert in their own right, and this knowledge must also be evaluated in the context of competing knowledges. Reactions to such conflict result from an evaluation of the information given and the sources providing that information, and range from changing behaviour to match the most recent information received (as with the many who resumed the diet after having been off it for a period of time as per their physicians' recommendations); to the response of one participant who said in regard to the changing advice on optimal blood levels, 'I'll just sit tight' (19 July 2005, interview), thus choosing to maintain a self-guided, but medically informed, status quo. Some people chose to ultimately discard medical recommendations in favour of their own expertise, as did one participant who did not perceive a difference in her functioning when she did not maintain the PKU diet. Her response of, '…well, to me, you know, there's no difference in how I feel…[*so*] I don't actually see a point to doing it…' (8 October 2005, interview) in a sense places her own knowledge of her body above the medical knowledge of her condition. (The medical view of course, suggests that the more noticeable and irreversible complications of treatment non-adherence would manifest themselves in subsequent years).

In such issues of treatment adherence, both the medical community and affected individuals are evaluating the concept of risks. Risk, however, as Douglas (1992: 46) has noted, is 'not a thing, it is a way of thinking, and a highly artificial contrivance at that' (Douglas 1992: 46). The recognition of risk is therefore far from universal, being instead an intensely situated phenomenon. This simple observation helps to make sense of some individuals' decisions not to adhere to the PKU diet. As Douglas and Wildavsky (1982) note, the ranking and prioritising which is so central to the process of risk assessment is a very subjective activity. One participant, for instance, said of the risk of future harm from being off-diet:

> …I'm overweight. There's diabetes in my family, there's lots of heart things in my family, there's various other things. There's lots of cancer in my family. I've got all these things kind of looming on my horizon anyway. It's just another possibility.

She therefore concluded:

> … I would say if *[adherence to dietary treatment*] makes a difference, it would not be a big difference. It would be a slight difference. And particularly with my other bits and pieces that I've got thrown into the mix as well…I think…that controlling PKU's not necessarily going to make a big difference, controlling blood levels is not necessarily going to make a big difference to my old age life (8 October 2005, interview).

The subjectivity and contextual dependency of decisions based on risk perception have been described by Lupton as 'competing knowledges about the world' (1999: 106), and one of the sites of primary competition is between expert (in this case, medical) knowledges of risk and lay knowledge (ie the knowledge belonging to people living with PKU). For example, Petersen and Lupton observe that '*[t*]he medical practitioner will tend to interpret data on risk through her or his own emotionally charged experience in working with individuals, having responsibility for their care and treatment, and being in the position of seeing patients die or avert death' (1996: 50). For these reasons, they argue, medical practitioners will tend to view risks as more severe or negative than will patients, and will expect a higher level of adherence to recommended treatment to avoid the risk. Lay people, on the other hand, 'may embrace official definitions of risk at some times, while at other times they may ignore or resist such definitions in pursuit of their own interests and goals' (*ibid.*). Their estimation of risk tends to be more contextual and localised, and more 'aware of diversity and change, than the universalising tendencies of expert knowledges' (Lupton 1999: 108). Lay people may draw from personal observation and the influence of other lay people (for example), in addition to expert information, in the generation of their risk knowledge. As a result of their tentative or ambivalent acceptance of medical estimates of risk, patients may express a lesser degree of adherence than would be expected by the medical community.

SOCIAL SPHERE

The participants in this project were very attuned to the social nature of food – even stating outright 'eating is a very social thing' (21 October 2005, interview). As a result, negotiation in the social sphere involved a great deal of impression management surrounding the stigma (in the sense described by Goffman) of not being able to meet the social expectations surrounding food consumption. The PKU diet (like other varieties of stigma) is a causal factor in the creation of an 'undesired differentness from what we had anticipated' (Goffman 1963: 15), which turns others away from the stigmatised individual. For many, this awareness of social difference begins at a very early age. For example, one interviewee related a childhood experience of having to refuse a chocolate cupcake at a friend's birthday party, remembering that '…even when you are little it feels like bad manners to say: 'I can't have that…' ' (21 October 2005, interview). Such feelings of difference could sometimes be mitigated by those whom Goffman refers to as the 'wise', i.e. individuals with some appreciation of the world of the stigmatised. One participant remembered:

> One of my neighbours was really, really good. Like, at Easter, because I couldn't eat Easter eggs, she used to get – you know those stockings way back in the `70s or something, those stockings what came in a…round mould that you take it apart – well, she used to take that, and she used to do a lot of icing, make wedding cakes and stuff, and she'd make it fancy with all the icing, and she'd leave it set, and that was my Easter egg (13 July 2005, interview).

The more common experience of living with PKU, however, seems to revolve around the necessity of repeated social indiscretions regarding food consumption. While participants varied in their perception of the degree of stigma relating to their condition (some feeling this quite acutely, and others not at all), most revealed parallels with the experience of a stigmatised individual. Goffman mentions for instance, that a stigmatised individual '…is likely to feel that he is 'on', having to be self-conscious and calculating about the impression he is making, to a degree and in areas of conduct which he assumes others are not' (Goffman 1963: 25). During the interviews with people with PKU, it was apparent that many were very impression-conscious, careful not to offend either with the refusal of food, or with breath that smells of the PKU supplement. The effect of such sensibilities is frequently to diminish their adherence to their treatment regimen. As follows: '…I went to my cousin's place for lunch…and she said do you eat fish? (*fish is not allowed on the* PKU *diet*) And I said, 'Yeah'.… I mean, she'd already bought it, and cooked it…' (18 October 2005, interview).

This is quite a striking example of what Goffman refers to as a 'formula' that stigmatised individuals are expected to follow when dealing with 'normals'. In essence, even though the stigmatised individual is the one burdened with the perceived shortcoming and consequent lack of social acceptance, they are expected to act in such a way as to benefit those without the stigma. For instance, Goffman notes that they become responsible for the avoidance of social situations where the stigma would be apparent, or where interaction between the stigmatised and nonstigmatised might be particularly uncomfortable. As an example, one participant was faced with a Christmas dinner for her antenatal group at a Japanese restaurant with extremely limited options for someone with PKU. Although she considered avoiding the situation entirely, for the sake of managing social relationships, she decided:

> I will probably still go for the social but I might eat before I go and I might just sit there and have a drink and you know, they have nice sorbets so I could have a sorbet for pudding so I might just say I will come for dessert and I will just arrive part way through their meal (21 October 2005, interview).

Her plan, outlined above, won out over the option of 'sitting there watching them all eat' for the reason that that would be 'embarrassing for them' (*ibid.*).

Specifically, individuals with PKU feel responsible for protecting people who do not have PKU from feeling awkward by not allowing their special diet to become too obtrusive. Theirs is also the responsibility for managing information regarding the disorder, and monitoring the way that speaking about PKU impacted on their personal relationships was a significant area of negotiations within the social sphere. Participants in this project represented a wide range of feelings on the issue of offering information about their condition. Some saw it as a 'good conversation starter' (19 July 2005, interview) and have become so accustomed to explaining themselves that it 'just all falls out' (21 October 2005, interview). Others, however, 'hate explaining to people what it is' (18 October 2005, interview).

Goffman (1963) has given some attention to issues of information control in the context of stigma. In addressing this matter, he distinguishes between the *discredited* individual, who '…assume[*s*] his difference is known about already or is evident on the spot…', and the *discreditable* individual, who '…assume[*s*] [*the stigma*] is neither known about by those present nor immediately perceivable by them…' (Goffman 1963: 14). If the person with PKU is not obviously displaying their dietary differences, and is in the company of those unaware of these differences, then as Goffman notes '[*t*]he issue is not that of managing tension generated during social contacts, but rather that of managing information about … [one's] failing. To display or not to display; to tell or not to tell; to let on or not to let on; to lie or not to lie; and in each case, to whom, how, when, and where' (Goffman 1963: 57). In constantly explaining her condition to people, one participant expressed that she felt she was '…undoing…or actually counteracting…what [*others*] might be thinking' (21 October 2005, interview). Understanding the negative social associations with rejecting food, she established that '…if you are turning something down all the time [*others will*] think you are rude, so…I am quite open about explaining [PKU] for that reason. I don't want people to think I am rude or a snob' (*ibid*.). Impressions can also be managed, not only in the conveying of information, but also in the manner in which that information is conveyed. One interviewee explained that her acquaintances who know of her condition are quite accepting of it, likely due to the fact that she does not 'make a big deal out of it' (18 October 2005, interview). By trivialising her own approach to the disorder, she felt that she successfully elicits a similarly casual response.

In another sense however, although the PKU diet does serve to distinguish the PKU community from the general public, it should also be noted that individuals with PKU are not the only people to differ from the 'norm' in terms of diet. It could be argued, then, that the dissimilarity that people with PKU experience as a result of their diet ironically places them in a community of others (constant dieters) who share a similar social distinction, and in many ways, a similar dietary experience. This sentiment was expressed by a number of participants, whereby the difference with others is then understood as a point of sameness.

For instance, when people diet, the goodness or badness of a food may not be merely symbolic, but may be perceived as a very real hazard or threat – certainly the case for people with PKU. '[*A*]ll diets work on the principle that food is dangerous' (Orbach 1978: 115). For the protection of the individual from the dangers that food poses, diets designate certain foods as legal and other foods as illegal. The moral world of foods for people with PKU is clearly and colourfully depicted in a helpful three-toned notebook distributed among the New Zealand PKU community, delineating between the 'legal' foods that (if eaten in normal quantities) can be consumed without restriction, the 'illegal' foods that should be avoided entirely, and the foods that are legal in moderation, to be regulated through the system of exchanges which was explained above (Nutrition Services: Auckland Hospital 2005).

The changing nature of medical recommendations for the PKU diet, which was raised by the majority of the interviewees for this project as an understandable source of some frustration, mimics the ways in which the dominant externally defined 'ideal' or 'correct' diet for health or weight loss also changes continually. Another shared experience with dieting for health or weight loss was the manner in which dieters find the externally defined standards of their diets to be quite unrealistic. In reference to energy reduction diets common among much of the female population of Western societies, Bordo suggests that '…total control [*over food*] is ultimately unsustainable…. The general point here is that 'the diet' is itself a precarious, unstable, self-defeating state for a body to be in…' (1993: 59). One of the results, according to Orbach, is that such externally defined diets have a basis that is 'far removed from the reality of women's day-to-day lives' (1978: 31). This was the case for many interviewees who, for example, found the time required to bake PKU bread and pastries to be unreasonable for people who also hold a job outside the home. This feeling can also be seen in the respondents' attitudes toward (and acceptance of) occasional non-adherence to the diet.

PERSONAL SPHERE

Yet another structuring element peppered throughout each interview was that of deprivation and, ultimately, of self-denial. The number of foods that participants 'love' but must not have, and the extreme limitation that this places on them appear to be central elements of the PKU experience and must be dealt with in one way or another. These restrictions seemed particularly salient for those who had spent much of their lives off-diet, but were now attempting to re-impose the dietary restrictions in their lives. For instance: 'I was able to give up…eggs, and meat, and fish, and cheese, and whatever else I had to give up. I liked corned beef quite a lot. I used to like eggs in a number of ways, you know – hard boiled, uh, scrambled, um, that sort of thing. Fish – I didn't mind fish. Hmm… I used to eat all those things' (9 July 2005, interview).

As mentioned previously, not only are certain types of food disallowed on the PKU diet, but also the amount of food consumed is restricted for many. For those with a more severe case of PKU, the counting of exchanges can be extremely restricting. As one participant stated, 'By the time you had two Weetbix for breakfast, you know, you hardly had room to pour milk on it and have a cup of tea, and you'd, you know, used up so many exchanges, sort of thing. It's quite horrendous, really' (18 October 2005, interview). Another participant attempted to stress this level of restriction by showing me her exchange book: '…I'm allowed one small potato, as an exchange…this is egg-sized. An egg-sized potato. Four of those would be four exchanges. Egg-sized potatoes. So just a small potato' (13 July 2005, interview).

Furthermore, people with PKU are deprived, not only of certain foods, but also more broadly of choice, freedom, and spontaneity in consumption. One interview felt that the hardest part of being on the PKU diet is 'not being able to eat freely'– '…you can't just sort of eat what you want' without thinking about the consequences (18 October 2005, interview). She longed for the 'freedom [*that*] normal people have got' (*ibid.*), pointing out that even such special occasions as dining out are somewhat spoiled due to this lack of freedom. The lack of variety permitted by the PKU diet was also a topic of much discussion. Variety, it seems, is necessary for many people to feel satisfaction with eating: '…you think you can just eat fruit and veggies. But you can't always keep doing that without thinking, 'I don't feel really satisfied' ' (9 July 2005, interview). The lack of variety in diet is perhaps felt more strongly in New Zealand than in other countries (like the US or Australia), as the supply of medical (low Phenylalanine) foods is limited due to the small size of the PKU population.

In addition to deprivation, many participants must negotiate incompatible lifestyle demands in their personal lives. As do members of the general population, individuals with PKU have strong feelings concerning the lifestyle they desire and for which they feel suited. For example, most of the women with PKU interviewed for this project (four of six) explicitly mentioned their disinterest in cooking and baking. As one participant so eloquently stated, 'I'm sorry, but women have gotten out of the kitchen' (9 July 2005, interview), or in another's words: '…do I want to live a life, or do I want to live in the kitchen?' (8 October 2005, interview). This dislike of cooking especially poses problems for women who attempt to prepare meals for their family in addition to themselves, essentially doubling their meal-preparation workload. This sentiment, therefore, represents a grave conflict between desired lifestyle and the demands of the PKU diet, as the medically recommended PKU diet reportedly requires substantial time spent in the kitchen (for example, baking bread and pastries using the low-protein flour). The result is a lifestyle out of step with the current world in which, as Mintz observes, 'more and more, people do not consume what they produce and do not produce what they consume, even much of their food' (1996: 10).

The time involved with meal preparation is not the only factor in competing lifestyle demands. In fact, the unpredictability of life in general is unhelpful in the quest to adapt to the PKU diet, when routine, for many, appears to be a key element of successful adherence. To this end, one participant explained that while attempting to maintain a very strict diet, 'If I changed something at work, or if the weather changed or something, my appetite changed, or, um something happened, like if in the weekend I wasn't at home controlling my meals and I was going out somewhere or something, it would all just completely fall apart' (8 October 2005, interview). Another interviewee asserted that a lifestyle truly compatible with the PKU diet would be quite sheltered, indeed: '…the best way for me to keep my blood levels where they want it is [*to*] live on my own, not work, stay at home, [*and*] never go anywhere for any temptations' (13 July 2005, interview).

A number of different strategies have been employed by PKU adults to negotiate the incorporation of the PKU diet into their personal lives. One such strategy is simply the development of tastes that coincide with the recommended diet. For instance, several participants mentioned their dislike of certain high-protein foods, such as chicken, egg, or cheese. This appears highly related to Bourdieu's notion of the 'taste of necessity', which, in his own words, serves as a form of 'adaptation to and consequently acceptance of' (1984: 372) one's circumstances. Alternatively, as mentioned above, habit and routine appear to play a role for many in adherence to the dietary restrictions. Yet another strategy may be found in PKU adults' attitudes toward food and eating, which tend to be more functional than experiential. Comments such as, '…you eat to live, you don't live to eat…' (21 October 2005, interview) express this well. Even more specifically, these people on the PKU diet think of food in relation to the physiological effects that it has on them personally, seeing food items in terms of exchanges or protein content, and therefore physical benefit or harm to themselves (9 July 2005, interview; 21 October 2005, interview). This can very much serve as a strategy to cope with the deprivation inherent in the PKU diet. By minimising the importance of pleasure in eating, the deficiencies in, for example, variety and tactile pleasure that the PKU diet possesses become less significant. As one participant related, '…food is very much just a sustenance thing, as opposed to a central focus. We focus on other things, I think, and get enjoyment from that' (9 July 2005, interview). Some participants also revealed complicated theories of acceptable dietary flexibility, allowing for certain types of transgressions (but not others), or outlining methods of 'making up' for previous lapses in dietary control.

THE ROLE OF THE NEGOTIATOR AND ITS IMPLICATIONS FOR THE MEDICAL ENCOUNTER

As a result of these competing demands and ambivalent social influences, most of the participants seemed quite independent in their self-regulation, knowledgeable of their needs, and capable of seeking out solutions. Thus, an approach of proactiveness toward medical support is evident from the interviews. One interviewee expressed this sentiment in terms of 'self-help', claiming to feel quite confident in her abilities to maintain her heath independently under most circumstances, and able to call on doctors and dieticians when she needs assistance (21 October 2005, interview). Medical professionals can best be of service in this type of relationship through enhancing their accessibility and approachability. They can also serve as providers of information, and several participants mentioned that more easily accessible information concerning current knowledge and research on PKU was needed. The more extensive provision of PKU-related information could possibly serve to bridge the gap between medical and self-expertise, and would undoubtedly aid individuals with PKU in their evaluative role of negotiation.

As demonstrated above, managing a PKU diet takes a great deal of thought and effort – from the evaluation of risk and contradicting information, to the management of relational complications caused by the diet, to the strategising of the inclusion of the diet in the personal sphere. Considering that this negotiation takes place in an atmosphere of independence, self-discipline, self-reliance, and proactiveness, PKU at times seems to be as much of a 'mental burden' (in the words of one participant) for those who suffer from it as a physical one (8 October 2005, interview). This participant's concept of the 'mental burden' has been quite useful in explaining and describing many of the stresses involved in the management of PKU in all three spheres explored in this paper. Time and time again, participants reaffirmed that PKU and the PKU diet is a constant consideration: 'It's just one of those things you always [*have*] to be thoughtful about' (9 July 2005, interview). One participant told the first author that PKU is 'on her mind' 'probably every day' (18 October 2005, interview). 'Thinking', therefore, becomes an intrinsic aspect of eating for people with PKU. '[*Other*] people…don't have to think, you know, they just eat. Whereas, I think when you have a diet like [*for* PKU], you kind of think as you eat, or think as you are about to eat' (21 October 2005, interview). Suggestions made to ease this difficulty include the preparation and provision of multiple PKU menus (and variations on these menus for those with different numbers of exchanges allowed per day) consisting of quick, economical foods, and cooking classes for PKU adults – or even better, for PKU children and adolescents in preparation for an adulthood of self-reliance. An increase in variety of low-phenylalanine medical foods, and especially convenience foods could also benefit PKU adults struggling with competing lifestyle demands.

This research clearly demonstrates through the above observations that the effects of PKU reach far beyond the physiological manifestations of the condition. According to the interviewees, the implications of PKU and its treatment are so numerous, apparently touching nearly every area of their lives, that PKU-related considerations must be dealt with continually: '…it's amazing where it crops up, you know, what you have to remember, what you have to allow for'; '…you've got to watch out, it crops up everywhere' (9 July 2005, interview).

CONCLUDING THOUGHTS

Most of the biomedically oriented research in the area of PKU has focused on treatment efficacy – exploring the physiological, behavioural, and mental outcomes of treatment and non-treatment at various stages of the life cycle. The approaches have been quantitative and positivist; looking, for example, at the relationship between depression and metabolic control, or assessing the quality of life of patients at differing stages of treatment (see Frank, Fitzgerald and Legge (in press) for a detailed discussion of this literature). This has meant that the social factors and consequences of 'compliance' or 'adherence' remain overlooked and also that key elements of the biomedical discussion are significantly under-theorised (such as the notions of 'compliance', 'adherence' and 'consumption') to the extent that the predictive and explanatory power of such research is severely curtailed from its very outset.

One remedy to this situation (and the path followed on this particular study) is to routinely apply two, complementary pools of expert knowledge which are already in existence to medical 'problems'. These are the extent social science literature which often provides a robust theoretical underpinning to the salient issues identified in the biomedical literature and also the experience near accounts from people living with the condition (usually gathered through ethnographic research). These expert knowledges are highly relevant to the goals of medicine but frequently poorly explored. The startling contrast provided within this study to the findings of the biomedical literature which stress the importance of social support to treatment adherence is a good example of the new perspectives to old problems which a hybrid knowledge base can produce. In our own study, people living with PKU routinely observed that 'other people' instead of providing social support, offered potent sources of reasons to stray 'off-diet' – a finding which is easily anticipated by anyone familiar with the social science literature on consumption, social power and identity formation but quite unexplored in the biomedical literature.

Our argument then has been that anthropology and the social sciences in general can provide this missing complexity in current biomedical explanations of medically defined problems such as 'adherence'. However, the successful application of this principle of incorporating social science ideas into medical problem solving is far more difficult to act upon that it is to envisage. The difficulties revolve around the issues of audience, textual construction and particularly the genre rules governing publication in the biomedical versus the social sciences. For anthropologists to contribute successfully to medical problem solving not only must we 'do' the research, we must develop a community of hybrid readers through whom the hybrid texts that we produce will circulate and finding such a cross disciplinary group of readers can be by far the most challenging aspect of the entire project.

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